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THE VERTEBRAL COLUMN AND TRUNK- DISORDERS
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The Vertebral Column and Trunk - Disorders

Deforming Diseases

Scoliosis

It might be said that to understand the nature of scoliosis, one must have an appreciation of Fryette's Laws. Fryette's Laws is a set of three laws pertaining to skeletal anatomy named after Harrison Fryette, D.O. The laws are defined as a set of guiding principles used by practitioners of osteopathic medicine to discriminate between dysfunctions in the axial skeleton. The first two laws solely apply to the lumbar and thoracic spinal regions, but the third applies to the entire vertebral column.

The three principles are:

1. **Principle I**: When the spine is in neutral, sidebending to one side will be accompanied by horizontal rotation to the opposite side. The involved group of vertebrae demonstrates a coupled relationship between sidebending and rotation. When the spine is neutral, side bending forces are applied to a group of typical vertebrae and the entire group will rotate toward the opposite side: the side of produced convexity. Extreme type I dysfunction is similar to scoliosis.

2. **Principle II**: When the spine is in a flexed or extended position (non-neutral), sidebending to one side will be accompanied by rotation to the same side. This law is observed in type II somatic dysfunction, where only one vertebral segment is restricted in motion and becomes much worse on flexion or extension. There will be rotation and sidebending in the same direction when this dysfunction is present.

3. **Principle III**: When motion is introduced in one plane it will modify (reduce) motion in the other two planes. The third principle sums up the other two laws by stating dysfunction in one plane will negatively affect all other planes of motion.

The first two laws were developed by Dr. Fryette in 1918, and the third was developed by C.R. Nelson, D.O in 1948.

In most cases, the cause of scoliosis is unknown (what doctors call idiopathic). The other of cases fall into two groups:

- **Non-structural** (functional): This type of scoliosis is a temporary condition when the spine is otherwise normal. The curvature occurs as the result of another problem. Examples include one leg being shorter than another, from muscle spasms due to injury, or from abdominal problems such as appendicitis.

- **Structural**: In this type of scoliosis, the spine is not normal. The curvature is caused by another disease process such as a birth defect, muscular dystrophy, metabolic diseases, and connective tissue disorders such as Marfan's syndrome.

Causes of scoliosis

Below are some of the possible causes of scoliosis:

- Neuromuscular conditions - these affect the nerves and muscles and include cerebral palsy and muscular dystrophy.
  - A primary muscle disorder has been postulated as a possible etiology of idiopathic scoliosis. The contractile proteins of platelets resemble those of skeletal muscle, and calmodulin is an important mediator of calcium-induced contractility. Kindsfater et al studied the level of platelet calmodulin in 27 patients with adolescent idiopathic scoliosis. Using a direct measurement technique, they showed that patients with a progressive curve (>10° progression) had statistically higher platelet calmodulin levels (3.83 ng/μg vs...
0.60 ng/μg). If these data are reproduced in larger studies, they hold the potential to allow clinicians to identify patients at higher risk of curve progression.

- **Congenital scoliosis (present at birth)** - this is rare and occurs because the bones in the spine developed abnormally when the foetus was growing inside the mother.
  - An elastic fibre system defect (abnormal fibrillin metabolism) has been offered as one potential aetiologic explanation for idiopathic scoliosis. Such abnormal connective tissue has not been found universally in patients with idiopathic scoliosis. No clear cause-and-effect relationship has been established. Further research in this area is clearly warranted.
  - Disorganized skeletal growth, probably with its root cause at a gene locus or group of loci, has been discussed as a possible etiologic explanation for idiopathic scoliosis. This theory is simply that a rather localized primary growth dysplasia leads to a cascading Hueter-Volkmann effect on a much larger portion of the spine. The Hueter-Volkmann principle states that compressive forces tend to stunt skeletal growth and that distractive forces tend to accelerate skeletal growth. A possible, yet unproven, association with such a growth disturbance is the osteopenia that has been identified in patients with idiopathic scoliosis.
  - Aronsson conducted a series of experiments exploring this mechanical modulation of growth. Using two different animal models (rats and calves), he showed that the force exerted by external ring fixators were quite capable of producing vertebral segment wedging akin to that seen in human idiopathic scoliosis. Correlation of his laboratory information with the clinical setting has drawn attention to the fact that wedging occurs both from the vertebral bodies themselves and from the disk spaces, with more thoracic wedging coming from the vertebral bodies. The asymmetric mechanical forces have also been associated with elevated synthetic activity in the convex side of scoliotic curves.
  - Bylski-Austrow and Wall led a group of Cincinnati Children's Hospital researchers who further analyzed the mechanical modulation of spinal growth. Using a porcine model, they successfully induced growth changes by means of an endoscopically implanted spinal staple. Within the context of 8 weeks' follow-up, they were able to create 35-40° of scoliotic curvature in growing pigs. Histologic analysis of vertebral specimens revealed increased paraphyseal density and disorganized chondrocyte development in the region of the staple blades.

- **Genes** - at least one gene is thought to be involved in scoliosis.
  - Genetic roots of the disease referred to as idiopathic scoliosis have been rather strongly suggested by several avenues of research. An X-linked inheritance pattern (with variable penetrance and heterogeneity) has been suggested by several authors. Studies of twins with scoliosis have pointed in a similar direction. More than 90% of monozygotic twins and more than 60% of dizygotic twins demonstrate concordance regarding their idiopathic scoliosis. Some evidence has also directed attention to portions of chromosomes 6, 10, and 18 as possible scoliosis-related loci.

- **Leg length** - if one leg is longer than the other, the individual may develop scoliosis.
- **Other causes** - bad posture, carrying backpacks or satchels, and some injuries.
On reflection, it can be said that the bones are the last thing to form during the embryonic and foetal stages of development. The bones are also the last thing to be left behind after we have died. It can easily be said that the bones are manifestations of any functional situation that has been present for a long time. The shape of the bones are defined by the shape of the original cartilaginous matrix, the ossification process and how that is modified by external factors; forces pulling on the developing skeleton, creating shape. One significant factor of this could be the underlying fascial pulling (not just the peri and epimysium). If such fascial pulling is persistent, then bony change will follow. Hence there is modification of skeletal alignment and the formation of facet alignment away from 'normal' becomes apparent. This is never the end result, though, as it can modify continually throughout life, often increasing in severity.

Scoliosis itself occurs when either flexion or extension is present, with ipsilateral rotation, especially during the growth years. This leads to facet alignment anomalies along the length of the spine. The alignment of the facets defining the type and even direction of movement at each individual level. When such changes occur in the thoracic spine, there can be a consequent decrease in thoracic function.

Idiopathic Scoliosis

Idiopathic scoliosis is the most common type of spinal deformity confronting orthopaedic surgeons. Its onset can be rather insidious, its progression relentless, and its end results deadly. Proper recognition and treatment of idiopathic scoliosis help to optimize patient outcomes. Once the disease is recognized, effective ways exist to treat it.

Scoliosis represents a disturbance of an otherwise well-organized 25-member intercalated series of spinal segments. It is, at times, grossly oversimplified as mere lateral deviation of the spine, when in reality, it is a complex three-dimensional (3D) deformity. In fact, some have used the term rotoscoliosis to help emphasize this very point. Two-dimensional (2D) imaging systems (plain radiographs) remain somewhat limiting, and scoliosis is commonly defined as greater than 10° of lateral deviation of the spine from its central axis.

In the past, terminology such as kyphoscoliosis was inappropriately used to describe certain patients with idiopathic scoliosis. Idiopathic scoliosis has a strong tendency to flatten the normal kyphosis of the thoracic spine. Winter taught that idiopathic scoliosis is a hypokyphotic disease. In most cases, diagnoses of kyphoscoliosis were clinical misinterpretations of the rib hump associated with an otherwise hypokyphotic thoracic spine. Idiopathic scoliosis may present as a true kyphoscoliosis, but this occurs relatively rarely.

James is credited with classifying idiopathic scoliosis according to the age of the patient at the time of diagnosis:

- Younger than 3 years have infantile idiopathic scoliosis
- Those aged 3-10 years have juvenile idiopathic scoliosis
- Those diagnosed when they are older than 10 years have adolescent idiopathic scoliosis

These age distinctions, though seemingly arbitrary, have prognostic significance. For instance, Robinson and McMaster reviewed 109 patients with juvenile idiopathic scoliosis and found that nearly 90% of curves progressed, and almost 70% of these patients went on to require surgery. These rates are much higher than the rates associated with other categories of idiopathic scoliosis. The real challenge is to predict which curves will progress significantly and which ones will not.
Historical viewpoint

Ancient Hindu religious literature (circa 3500-1800 BCE) describes the treatment of spinal deformity rather clearly. The story is told of a woman who was "deformed in three places" and how Lord Krishna straightened her back. This was accomplished by pressing down on her feet and pulling up on her chin. The orthopaedic trappings of the story are unmistakable, including excellent immediate posttreatment results and no long-term follow-up.

Hippocrates (circa 400 BCE) stated, "There are many varieties of curvature of the spine even in persons who are in good health; for it takes place from natural conformation and from habit." He also stated that "lateral curvatures also occur, the proximate cause of which is the attitudes in which these patients lie." The postural and muscular theory of scoliosis thus stated has persisted for thousands of years and remains firmly embraced by some.

Hippocratic scoliosis treatment methods focused primarily on spinal manipulation and traction. He used an elaborate traction table called the scennum. Medical practitioners used slight variations of the Hippocratic scennum well into the 1500s. Another treatment approach that Hippocrates discussed involved attempting to diminish spinal deformity with a method called succussion. This involved strapping the patient (often upside down) to a ladder, which was then hoisted into the air and dropped from a height. Hippocrates thought that this method was occasionally useful, but it was largely performed by charlatans to impress the public.

Ambroise Pare, the "most celebrated surgeon of the Renaissance," is recognized as the first physician to treat scoliosis with a brace. He also recognized that once a patient with scoliosis had reached maturity, bracing was not useful. Pare's orthosis consisted of a metal corset (fashioned in a village smithy setting) with many holes in it to help diminish its significant weight. The record also makes it quite clear that Pare espoused the postural theory of scoliosis.

Nicholas Andry was a French paediatrician who hated the brutal barber surgeons of his day. At the age of 83 (a year before his death) he wrote a short book entitled Orthopaedia. Thus, in 1741 this name combined the root words for "straight" (orthos) and "child" (pais) to create the name still used for the broad musculoskeletal field, orthopaedics.

Andry believed that scoliosis was caused by asymmetric muscle tightness and, thus, helped foster the French belief in "convulsive muscular contraction" as the cause of spinal deformity. Andry stated, "It is well worth while to remark that the crookedness of the spine does not always proceed from a fault of the spine itself, but is sometimes owing to muscles of the forepart of the body being too short, whereby the spine is rendered crooked, just in the same manner as a bow is made more crooked by tying its cord tighter." Andry used rest, suspension, postural approaches, and padded corsets in his treatment of scoliosis.

Jacques Mathieu Delpech was a successful and skilled surgeon, yet he focused a great deal of his attention on nonsurgical approaches to orthopaedic problems. For the treatment of scoliosis, Delpech devised graded exercises for strengthening muscles of the trunk in the belief that the deformity was due to a weak axial musculature. This belief was almost certainly due to the influence of Andry. Delpech also used stretching and traction techniques but did not believe in braces.

An important event of the 1800s was the advent of surgical treatment of scoliosis by the French orthopaedic surgeon Jules Guerin. He was very enthusiastic about subcutaneous tenotomy and myotomy and first reported their use in his scoliosis patients in 1839. When he later published the results of treatment of 1349 patients with this technique, tremendous controversy was ignited. Guerin's harshest critic was Joseph Malgaigne, who described
Guerin’s work as “some orthopaedic illusion.” This led to one of the most famous orthopaedic lawsuits in history: Guerin versus Malgaigne. This defamation trial ended in Malgaigne’s favour and helped to establish an important precedent for open criticism of scientific papers.

Another important tool in the treatment of scoliosis was the plaster body jacket (i.e., body cast). The American orthopaedic surgeon Lewis Sayre popularized its use in the mid-1800s. Sayre’s technique involved a large tripod that allowed the patient to be suspended while the corrective plaster cast was applied. Sayre was said to be “a brusque, forceful and therefore controversial personality” but also “an eloquent speaker” who toured internationally demonstrating his casting techniques.

The early 1900s saw what was arguably the most important advance in scoliosis treatment in more than 3000 years - posterior spinal fusion. Russell Hibbs first performed his “fusion operation” for tuberculous spinal deformity in 1911, but by 1914 he also was applying his technique to patients with scoliosis. The Hibbs approach focused on achieving maximum deformity correction via a variety of plaster jackets before surgery.

Further refinement in surgical technique and instrumentation has led to the greater than 50% correction and single-digit pseudarthrosis rates to which contemporary orthopaedists have become accustomed.

The vast majority of patients with idiopathic scoliosis initially present because of a perceived deformity. This may be patient or family perception of asymmetry about the shoulders, waist, or rib cage. A primary care physician or school-screening nurse may perceive similar findings. The Adams forward-bending test (in conjunction with the use of a scoliometer) has been found to be an effective screening tool.

Highlights of the patient's history include information relative to other family members with spinal deformity, assessment of physiologic maturity (e.g., menarche), and presence or absence of pain.

Traditionally, scoliosis has been described as a non-painful condition. Ramirez et al from the Texas Scottish Rite Hospital studied more than 2400 patients with scoliosis and found that a full 23% (560 of 2442 patients) had back pain at the time of presentation. An underlying pathologic condition was identified in 9% (48 of 560) of the patients with back pain, including mainly spondylolysis and spondylolisthesis but also intraspinal tumour in one instance. Thus, it would seem that pain is not associated with scoliosis as rarely as was previously thought.

Epidemiology

- Age - scoliosis signs and symptoms often start during a growth spurt that occurs just before puberty
  - Stirling et al studied almost 16,000 patients aged 6-14 years in England and found the point prevalence of idiopathic scoliosis was highest (1.2%) in patients aged 12-14 years. This has helped reiterate the idea that the focus of screening efforts should be on children in this age group.
- Gender - females have a higher risk
  - Scoliosis has been suggested to develop more frequently in children born to mothers who are aged 27 years or older. One might hypothesize that gene fragility might be involved (e.g., higher rate of infants with Down syndrome born to older mothers). The precise explanation as to why this might be the case has not been elucidated. In addition to this, no other authors have duplicated these results.
As mentioned previously, most patients with idiopathic scoliosis are female, and the vast majority of research has focused on females. One of the only articles written on idiopathic scoliosis in males is that by Karol et al, from the Texas Scottish Rite Hospital. These authors showed that boys with scoliosis are at risk for curve progression for a longer period than girls. They also suggested that efforts to screen for boys with scoliosis should be performed a little later than similar screenings for girls.

- Genetics - people with scoliosis are more likely to have close relatives with the same condition than people without scoliosis

**Prognosis**

Clinical outcomes following treatment of idiopathic scoliosis are strongly linked to curve magnitude. Unrealistic presurgical expectations have been shown to correlate with a decreased likelihood of postsurgical satisfaction.

Studies have shown that reoperation for implant/fusions have been necessary for between 3.9% - 9.2% of patients, 4% for loosening of implants or even their removal.

**Imaging Studies**

**Radiography**

The thoracic curve patterns found in adolescent idiopathic scoliosis are still most commonly classified according to the King classification system. Significant questions have been raised regarding the reliability and reproducibility of this system. In addition to this, the King classification alone (in its original form) does not allow comprehensive curve classification (e.g., lumbar and thoracolumbar curve patterns).

*Figure 1 Scoliosis - A-P X-ray*

**Computed Tomography**

The scoliotic deformity can be visualized on CT scans of the thorax and abdomen (see the images below). Associated lesions, such as osteoid osteoma, osteoblastoma, infection, tumours, disk prolapse, and costovertebral dislocation, can be found.
CT scanning with sagittal and coronal reconstructions can provide all the information that a plain radiograph provides (see the images below). With CT, 3D reconstructions are useful in assessing segmentation abnormalities. CT can also be used to assess the true extent of rotation and rib deformities. It plays an important role in evaluating postoperative complications.

*Figure 2 Scoliosis - Coronal CT scan showing several hemivertebrae*

Idiopathic scoliosis is diagnosed only after underlying structural conditions such as like hemivertebrae are excluded.

*Figure 3 Scoliosis - 3D reconstructed CT of same patient, showing hemivertebrae*

*Figure 4 Scoliosis - CT is good for assessing rotation of the vertebrae*

*Figure 5 Scoliosis - CT scan illustrates extent of rotation*

With modern multidetector-row CT scanners, thin sections of the whole body can be obtained within a few seconds and reconstructed in any plane: sagittal, coronal, oblique, or axial. Even 3D reconstruction with shaded-surface display or volume rendering is possible.

The role of MRI is controversial. Although some institutes prefer to perform routine preoperative MRI in all patients, studies have shown that such an approach does not provide clinically significant results. MRI is useful for diagnosing associated spinal and neurologic lesions. Small tumours and infections can be localized by using MRI.
Limitations of techniques

The main limitation of radiography is the radiation dose. The risk of carcinogenesis is increased because of the repeated examinations done to monitor curve progression. This risk can be reduced with the judicious use of radiography and proper protection techniques.

Radiography is less sensitive than bone scanning and MRI because tumours or infections are apparent only after 50% of the bone is destroyed. Radiographs cannot be used to assess abnormalities of the spinal cord.

CT scanning is not routinely indicated, but it is a good method for assessing rotation and segmentation abnormalities. Radiography can provide all of the information needed. MRI is not cost-effective, and it is not a good screening tool because its yield in depicting important clinical abnormalities that change management is minimal.

Histologic Findings

Scoliosis is clearly a disease that is strongly influenced by, if not completely rooted in, spinal growth. It has even been referred to by some as "an unsynchronized growth."

In a study by Hsu et al (studying muscle biopsies), 68% of the patients demonstrated abnormalities in muscle fibre distribution. The abnormalities were similar on the convex and...
concave sides, the most notable being a reversal of the normal type 2 fibre ratio, so that type 2A fibers predominated over type 2B fibers in the study subjects. These changes are similar to those seen in endurance training and might be due to the extra work of trying to maintain posture in the setting of scoliosis.

**Infantile idiopathic scoliosis**

**Scoliosis signs and symptoms in babies**

- A bulge on one side of the chest
- The baby might consistently lie curved to one side
- In more severe cases, the heart and lungs may not work properly, and the patient may experience shortness of breath and chest pain

Although defined by a seemingly arbitrary age limit (<3 years at the time of diagnosis), infantile idiopathic scoliosis demonstrates marked differences that distinguish it from the other two categories of idiopathic scoliosis.

Infantile idiopathic scoliosis is the only type of idiopathic scoliosis whose most common curve pattern is left thoracic. It is the only type of scoliosis that is more common in boys. It is more common in European patients or those of immediate European descent. In the past, infantile idiopathic scoliosis may have constituted up to 41% of all idiopathic scoliosis cases in parts of Europe, but the current rate would appear to be closer to 4%. This is still dramatically higher than the estimated 0.5% rate in North America.

Infantile idiopathic scoliosis is also the only type of idiopathic scoliosis with any significant reputation for spontaneous resolution. Reported spontaneous resolution rates are in the range of 20-92%. Ceballos et al studied 113 Spanish patients with infantile idiopathic scoliosis. They found a 92% rate of associated plagiocephaly and an almost 25% rate of congenital hip dysplasia. In addition, they found that nearly 74% of their patients’ curves were of the resolving variety (mainly left thoracic curves) and the other 26% were progressive curves (mainly double primary type curves).
Prediction of curve progression in infantile idiopathic scoliosis has been tied to assessment of the rib vertebral angle difference (RVAD) originally described by Mehta in 1972. As described by Mehta, this measurement is carried out at the apical vertebra of the curve. In instances in which the curves resolved spontaneously, the RVAD was less than 20° in about 80% of cases, and in those instances in which the curves were progressive, the RVAD exceeded 20° in about 80% of cases. Other authors have confirmed the prognostic value of the RVAD, as well as its reliable application.

**Treatments for scoliosis**

The majority of children with scoliosis have mild curves and don't need treatment. In such cases, the doctor will recommend regular follow-ups every 4 to 6 months to monitor the curve of the spine in clinic and periodically with X-rays.

The following factors will be considered by the doctor when deciding on treatment:

- Gender - females are more likely than males to have scoliosis that gradually gets worse.
- Severity of the curve - the larger the curve, the greater the risk of it worsening over time. S-shaped curves, also called "double curves," tend to get worse over time. C-shaped curves are less likely to worsen.
- Curve position - if a curve is located in the center part of the spine, it is more likely to get worse compared with curves in the lower or upper section.
- Bone maturity - the risk of the curve worsening is much lower if the patient's bones have stopped growing. Braces are more effective while bones are still growing.

**Medical Therapy**

Nonoperative management consists of either simple observation or orthosis use. Observation is watchful waiting with appropriate intermittent radiographs to check for the presence or absence of curve progression. Orthosis use for scoliosis is discussed extensively below.

No other treatments, including electrical muscle stimulation, physical therapy, spinal manipulation, and nutritional therapies, have been shown to be effective for managing the spinal deformity associated with idiopathic scoliosis. The lack of demonstrated effectiveness in this context means either that scientifically valid studies have been done that do not support the treatment or that no such studies have yet been published that would allow an evidence-based evaluation.

The first widely used scoliosis brace with proven effectiveness was the Milwaukee brace. This brace was developed by Walter Blount and Albert Schmitt and introduced at a meeting of the American Academy of Orthopaedic Surgeons in 1946. It was originally designed to be used as part of the surgical treatment of scoliosis and only later evolved into a standalone non-operative treatment. Some authors have shown that an average curve correction of 20% in the brace (Milwaukee brace) is associated with bracing success.

Rowe et al performed a meta-analysis aimed at evaluating the efficacy of non-operative treatments for idiopathic scoliosis. They calculated the weighted mean proportion of success for three non-operative treatments: observation, electrical stimulation, and bracing. They were able to successfully combine data on 1910 patients from 20 different studies for purposes of meta-analysis and reported the following main results:

- Observation, 49% success rate
- Electrical stimulation, 39% success rate
• Bracing 8 hr/day, 60% success rate
• Bracing 16 hr/day, 62% success rate
• Bracing 23 hr/day, 93% success rate

In a prospective multicentre study from the Scoliosis Research Society, Nachemson et al found brace treatment (an underarm plastic brace worn for at least 16 hr/day) to be successful 74% of the time (95% confidence interval [CI], 52-84%). Some authors have not been able to identify a major difference between full-time bracing (23 hr/day) and part-time bracing (12-16 hr/day).

The psychological stress associated with scoliosis has been documented, and this does not improve compliance with brace wear. MacLean et al from Vanderbilt studied 31 adolescent and preadolescent females who were undergoing part-time brace treatment for their idiopathic scoliosis. Part-time bracing was defined as 13-16 hr/day. The investigators noted that 84% of patients described the initial period of bracing in "stressful terms" and experienced lower levels of self-esteem. A reassuring finding was that no overt psychopathology was identified in this study.

Compliance with prescribed brace-wear regimens has been shown to be poor. DiRaimondo and Green found that on average, patients only wore their braces 65% of the prescribed amount of time. Patients prescribed part-time (16 hr/day) bracing actually demonstrated worse compliance (58%) than those prescribed full-time (24 hr/day) bracing (71%). Overall, only 15% of patients demonstrated a highly compliant (≥90%) brace-wear routine.

Questions have also been raised regarding the consistency of strap tension in TLSO bracing. Using an instrumented load cell to measure strap tension, Aubin et al studied 34 of their patients with braces in Quebec. They found marked variability in tension, with the greatest change occurring while patients were recumbent.

In part because of the aforementioned psychological and brace-wear compliance issues, new approaches to bracing are being developed. One such approach, developed by Coillard and Rivard of the St Justine Hospital in Montreal, is a dynamic bracing method known as the SpineCor Brace or as the St Justine Brace. It involves elastic straps that are anchored on a pelvic corset, and based on curve morphology, these straps are tensioned to exert corrective forces. The brace is a radical departure from traditional plastic and metal orthoses.

Early results with the St Justine Brace are encouraging, with success rates comparable to those of traditional bracing. Continued follow-up of their growing international cohort of patients is necessary. A study by Gutman et al found the SpineCor brace to be less effective than the Boston brace for treatment of adolescent idiopathic scoliosis.

TREATMENT INDICATIONS FOR INFANTILE SCOLIOSIS

Casting

Casting instead of bracing is sometimes used for infantile scoliosis to help the infant's spine go back to its normal position as it grows. This can be done with a cast made of plaster of Paris.

The cast is attached to the outside of the patient's body and will be worn at all times. Because the infant is growing rapidly, the cast is changed regularly.
Non-operative treatment of progressive infantile idiopathic scoliosis predominates and may involve the use of conventional thoracolumbosacral orthosis (TLSO)-type braces, Milwaukee-type braces, and even intermittent Risser casting. Some have questioned the value of bracing in infantile idiopathic scoliosis and have stated that "a curve that resolves in a brace would probably have resolved without treatment."

**Braces**

If the patient has moderate scoliosis and the bones are still growing, the doctor may recommend a brace. This will prevent further curvature, but will not cure or reverse it. Braces are usually worn all the time, even at night. The more hours per day the patient wears the brace, the more effective it tends to be.

The brace does not normally restrict what the child can do. If the child wishes to take part in physical activity, the braces can be taken off (check with the doctor).

When the bones stop growing, braces are no longer used. There are two types of braces:

- Thoracolumbosacral orthosis (TLSO) - the TLSO is made of plastic and designed to fit neatly around the body's curves. It is not usually visible under clothing.

- Milwaukee brace - this is a full-torso brace and has a neck ring with rests for the chin and the back of the head. This type of brace is only used when the TLSO is not possible or not effective.

One study found that when bracing is used on 10- to 15-year-olds with idiopathic scoliosis, it reduces the risk of the condition getting worse or needing surgery.
Surgery (spinal fusion)
In severe cases, scoliosis can progress over time. In these cases, the physician may recommend spinal fusion. This surgery reduces the curve of the spine and stops it from getting worse.

Two or more vertebrae (spine bones) are connected with new bone grafts. Sometimes, metal rods, hooks, screws, or wires are used to hold a part of the spine straight while the bone heals.

The operation lasts from 4 to 8 hours. After surgery, the child is transferred to an ICU (intensive care unit) where they will be given intravenous fluid and pain relief. In most cases, the child will leave the ICU within 24 hours, but may have to remain in hospital for a week to 10 days.

Children can usually go back to school after 4 to 6 weeks, and can take part in sports roughly 1 year after surgery. In some cases, a back brace is needed to support the spine for about 6 months.

The patient will need to return to the hospital every 6 months to have the rods lengthened - this is usually an outpatient procedure, so the patient does not spend the night. The rods will be surgically removed when the spine has grown.

Preoperative considerations
Preoperative evaluation focuses on specifics of curve location, magnitude, and flexibility. These parameters are used in conjunction with patient maturity factors to determine optimal treatment choice, but definitive studies are not yet available that dictate specific surgical tactics. However, the scoliosis surgeon is aided by commonly applied clinical guidelines that have evolved over time. The goal is always to fuse as little of the spine as possible while adequately treating existing major curvature.

For a thoracic curve (with adequate flexibility) without any significant associated lumbar curvature, the most common surgical approach has not changed since the days of Paul Harrington: posterior spinal fusion with instrumentation. Surgeons may choose from a diverse array of anchors to secure large-diameter rods (usually in the 0.25-in. range) to the spine. These anchors include sublaminar hooks, pedicle hooks, transverse process hooks, sublaminar wires (Luque wires), spinous process wires (Drummond wires), and pedicle screws.

Some surgeons have advocated anterior spinal fusion and instrumentation for such isolated thoracic curves. These have included both open (thoracotomy) and limited-incision (thoracoscopic) techniques.

When the primary problem is a large, stiff thoracic curve (usually not bending less than 50°), a different surgical tactic is usually undertaken in which an anterior release (usually including discectomy and bone grafting) is performed prior to posterior spinal fusion and instrumentation. Anterior spinal fusion and instrumentation has also been advocated in this situation, provided the patient does not have excessive kyphosis associated with a large thoracic curve.

Large curve patterns that include both thoracic and lumbar deformity continue to challenge scoliosis surgeons. If adequate flexibility and balancing of the lumbar spine is possible, then selective fusion of the thoracic curve is possible. When this is not the case, extensive fusion (at times down to the fourth lumbar segment) may become necessary.
The Scoliosis Research Society has a reasonably specific definition of thoracolumbar scoliosis: a curve whose apex lies at the body of T-12 or L-1 or at the T12-L1 interspace. These curves are most commonly left-sided curves, and they present one of the most common scenarios in which anterior spinal fusion and instrumentation is utilized.

Risks - the doctor will only recommend spinal fusion if the benefits are thought to outweigh the risks. The risks include:

- **Rod displacement** - a rod may move from its correct position. This happens in about 1 in every 20 cases of spinal fusion. Although not uncomfortable, the patient may need further surgery.

- **Pseudarthrosis** - one of the bones used to fuse the spine into place does not stick properly. This occurs in 1-5 percent of cases. Some patients may experience mild discomfort, and the spine will not be corrected as successfully. Further surgery may be needed.

- **Infection** - approximately 1-2 percent of spinal fusion patients develop an infection after the operation. This is usually treated with medication (antibiotics).

- **Nerve damage** - occurs in about 1 or 2 surgeries per 1,000. Damage occurs to the nerves of the spine. Results can range from mild, with just numbness in one or both legs, to paraplegia (loss of all lower bodily functions). A neurosurgeon may be present for scoliosis surgery.

If surgical treatment becomes necessary, anterior release and fusion followed by posterior spinal fusion with instrumentation is considered to be the functional treatment. Every effort should be made to delay such surgical intervention as long as possible to optimize spinal growth, but relentless curve progression should not be accepted or tolerated while some arbitrary chronologic age is awaited.

Although convex spinal epiphysiodesis (which has been shown to be quite effective in the management of congenital scoliosis) is intuitively attractive, it has not been shown to be as reliable in the setting of infantile idiopathic scoliosis. Addition of some type of posterior instrumentation may improve the results of epiphysiodesis.

A treatment outline for infantile idiopathic scoliosis may be as follows:

- Curves less than 25° with an RVAD less than 20° are preferentially observed and monitored with spinal radiographs at regular intervals.

- Curves exceeding these parameters are typically braced, with some consideration given to the value of intermittent Risser casting.

- Surgery is considered for curves not adequately controlled with non-operative measures.

**Juvenile idiopathic scoliosis**

*Scoliosis signs and symptoms in infants*

- The head is slightly off centre.

- The ribcage is not symmetrical - the ribs may be at different heights.

- One hip is more prominent than the other.

- Clothes do not hang properly.

- One shoulder, or shoulder blade, is higher than the other.

- The individual may lean to one side.

- Uneven leg lengths.
Juvenile idiopathic scoliosis most closely mimics the epidemiology and demographics of the adolescent version of the disease. It is more common in females, and its most common curve pattern is a right thoracic curve. In fact, given its demographic similarities, high rate of progression, and need for surgery, juvenile idiopathic scoliosis might be considered to be a malignant subtype of adolescent idiopathic scoliosis.

Robinson and McMaster studied 109 patients with juvenile idiopathic scoliosis in Scotland and found that 95% (104 of 109 patients) demonstrated curve progression and 64% (70 of 109 patients) progressed to require a spinal fusion. This spinal fusion rate is similar to that reported by James 15 years earlier.

A study from Washington University found a 50% rate of neural axis abnormalities in young children (<10 years) with idiopathic scoliosis. These findings included Chiari type I malformations and dural ectasia. At least one case report also exists in which a spinal intraosseous arteriovenous malformation was found in association with juvenile scoliosis.

Treatment indications

One potential treatment algorithm for juvenile idiopathic scoliosis is as follows:

- Observation for curves less than 25° with follow-up radiographs at regular intervals
- Bracing for curves that range from 25° to 40° and at least consideration of bracing (based on curve flexibility) for curves from 40° to 50°
- Bracing for smaller curves that demonstrate rapid progression to the 20-25° range
- Surgical intervention for inflexible curves that exceed 40° or virtually any curve that exceeds 50°

Bracing and casting may be used outside the above-mentioned parameters in an effort to help control a large curve in a young child for whom the surgeon is attempting to optimize spinal growth. Similar recommendations exist regarding the value of MRI in juvenile idiopathic scoliosis due to a significant rate of neural axis abnormalities.

Figure 11 Scoliosis - Preoperative thoracic scoliosis
Adolescent idiopathic scoliosis

Adolescent idiopathic scoliosis is the most common type of idiopathic scoliosis and the most common type of scoliosis overall. Progressive curvature may be predicted by a combination of physiologic and skeletal maturity factors and curve magnitude. Small curves in more mature patients have a substantially lower risk of progression (~2%) than larger curves in more immature patients, in whom the risk is much higher (approaching or exceeding 70%).

Contraindications for scoliosis correction

Few, if any, absolute contraindications exist regarding scoliosis care, just as few, if any, absolute indications for intervention exist. Accepted contraindications for bracing include skeletal maturity and excessive curve magnitude. Thoracic lordosis and certain curve patterns such as double thoracic curves also have been offered as at least relative contraindications to bracing.

The main contraindication to posterior scoliosis surgery would be medical instability and inability to survive surgery. Anterior scoliosis surgery would also be contraindicated in these patients, as well as in those with a precarious pulmonary status.

Surgical Therapy

Even in the setting of adequate correction and solid fusion, up to 38% of patients still have occasional back pain.

The primary goal of scoliosis surgery is to achieve a solid bony fusion. The surgical technique used to achieve such an arthrodesis is vastly more important than the instrumentation system that the surgeon needs to use, if any.

Anterior approaches to this area of the spine were pioneered by Hodgson (Hong Kong), Dwyer (Australia), and Zielke (Germany). Current approaches represent further refinement of these original techniques, such as modern large rod-and-screw systems and the John Hall short anterior segment overcorrection technique. The value of such techniques lies in their...
ability to powerfully correct large thoracolumbar curvatures while minimizing fused segments within the lumbar spine.

There is little debate regarding the fixation of the rods used for anterior instrumentation. Large bone screws are almost always the anchor of choice. For posterior instrumentation procedures, the surgeon has more options. Multiple hooks are the most commonly used anchors. They offer simplicity, strength, and near complete visualization during insertion. Their main drawbacks relate to size mismatch between hooks and associated bony elements, as well as the absence of appropriate hook sites (such as might be the case in myelomeningocele, tumour cases, or revision surgeries).

Sublaminar wires offer the power of segmental fixation and firm bony purchase, but with the drawback of possible dural and/or spinal cord trauma. As a result, either very selective use of or no use at all of sublaminar wires is usually the case in the setting of idiopathic scoliosis. A reasonable compromise was achieved when Drummond introduced his spinous process wires (also known as Wisconsin wires). These devices still offer the power of segmental fixation with virtually none of the nerve injury risks of sublaminar wires.

Pedicle screws have also become a popular anchor for the rods used in posterior scoliosis fusion procedures. They offer the potential advantage of increased strength (and possibly power of correction) while at the same time introducing added insertion-technique complexity and different neurologic complication risks. A very real and major increase in the overall cost of instrumentation constructs that include many pedicle screws is the case when they are compared to similar constructs that may include hooks and wires.

At this time, the available evidence in favour of a commensurate improvement in clinical outcomes is not sufficiently conclusive to support routine use of such pedicle screw constructs in the treatment of idiopathic scoliosis.

Pulmonary function testing is commonly used in the preoperative evaluation of patients with idiopathic scoliosis who are slated to undergo surgery. Such testing may influence the surgeon’s enthusiasm for related procedures, such as costoplasty (thoracoplasty). Pulmonary function testing may also uncover previously unrecognized tobacco use (an independent risk factor for pseudarthrosis) or undiagnosed (subclinical) pulmonary disease.

Postoperative radiographic assessment
In the immediate postoperative period, chest radiographs are used to assess for complications. Frontal and lateral spinal images are required to assess the postoperative spine. The degree of correction of the curve can also be evaluated.

Recognized complications are damage to the rods or wires, dislocation or migration of the rods, loosening of a device, spondylolysis, pseudoarthrosis, aortic aneurysm, retroperitoneal fibrosis, gastric volvulus, and progression of the deformity (crankshaft phenomenon). This last complication occurs when surgery is performed before maturity (Risser index < 1 and age < 10 y). The unfused vertebrae and portions of the vertebrae not included in the fusion continue to grow, producing progressive deformity. This effect is marked in posterior fusion, in which the anterior portions of vertebrae continue to grow and produce a new curve.
Follow-up assessment should be routinely performed until the patient reaches maturity.

**Scheuermann’s disease**

Scheuermann’s disease is a self-limiting skeletal disorder of childhood. It is also known as **Scheuermann’s kyphosis**, as it results in kyphosis. It is also known as: Calvé disease, and idiopathic juvenile kyphosis of the spine.

Scheuermann’s disease describes a condition where the vertebrae grow unevenly with respect to the sagittal plane; that is, the anterior angle is often greater than the posterior. This uneven growth results in the signature “wedging” shape of the vertebrae, causing kyphosis.

It is found mostly in teenagers and presents a significantly worse deformity than postural kyphosis. Patients suffering with Scheuermann’s kyphosis cannot consciously correct their posture. The apex of their curve, located in the thoracic vertebrae, is quite rigid. This rigidity is notorious for causing lower and mid-level back and neck pain, which can be severe and disabling. The sufferer may feel pain at the apex of the curve, which can be aggravated by physical activity and by long periods of standing or sitting. In addition to the pain associated with Scheuermann's disease, many sufferers of the disorder have loss of vertebral height, and depending on where the apex of the curve is, may have a visual 'hunchback' or 'roundback'. It has been reported that curves in the lower thoracic region cause more pain, whereas curves in the upper region present a more visual deformity.

The seventh and tenth thoracic vertebrae are most commonly affected. It causes backache and spinal curvature. In very serious cases it may result in internal problems and spinal cord damage, but these cases are extremely rare. The curvature of the back can put pressure on internal organs, wearing them out more quickly than the natural aging process; surgical procedures are almost always recommended in this case.

Along with this wedging of the vertebra there is also a change to the interface between the disc and the vertebra called endplate irregularities. Some of the disc then pushes into the vertebra and these are called **Schmorl’s nodes** and are typically seen on an X-Ray.
These Schmorl's nodes are present for life but are do not appear to cause any problems in the future. People may have an X-Ray when they are older for an unrelated condition and find that they have Schmorl's nodes but have never experienced back pain.

Its causes are unknown, though it isn't unreasonable to see tension patterns within the chest pulling the spine into a kyphosis, this being particularly significant during the growth years. The tension pattern pulling the thoracic spine into a kyphosis will also contribute to the deformation of the thoracic vertebrae and their consequent wedge shape.

The main focus of any therapy is to help maintain mobility of the thoracic region.

**Pectus Carinatum**

Pectus carinatum (i.e., carinatum or keel-shaped deformity of the chest) is a term used to describe a spectrum of protrusion abnormalities of the anterior chest wall (see the image below).

The deformity may be classified as either chondrogladiolar or chondromanubrial, depending on the site of greatest prominence. Lateral deformities are also possible.

Hippocrates described the carinatum deformity as a "sharply pointed chest" and reported that patients became "affected with difficulty breathing." Symptomatic patients report dyspnoea and decreased endurance. Some develop rigidity of the chest wall with decreased lung compliance, progressive emphysema, and increased frequency of respiratory tract infections. Many affected patients have no physical complaints; however, concerns about body image have been associated with low self-esteem and a decreased mental quality of life. Cosmetic concerns can be significant factors in opting for correction. Barrel chest deformities with increased anteroposterior (AP) chest diameters can be seen in obstructive forms of chronic pulmonary disease, such as cystic fibrosis and untreated or poorly controlled asthma.

**Pathophysiology**

Until recently, most cases of pectus carinatum deformity were thought to be asymptomatic. However, little is known about the cardiopulmonary function. In 1989, Derveaux reported a series of patients with no significant preoperative or postoperative respiratory compromise. However, some patients develop a rigid chest wall, in which the AP diameter is almost fixed in full inspiration. In these patients, respiratory efforts are less efficient. Vital capacity is reduced, and residual air is increased. Alveolar hypoventilation may ensue, with arterial hypoxemia and the development of cor pulmonale. As the lungs lose compliance, incidence of emphysema and frequency of infection are increased. Most recently,
Fonkalsrud (2008) reported his personal experience of 260 patients, all of whom were symptomatic. Symptoms that were reported included dyspnoea, exertional tachypnoea, and reduced endurance.

In 1990, Lakovlev and colleagues studied the cardiac functions of 70 patients with pectus carinatum deformity. Mitral valve prolapse was identified in 97%. Rhythm disturbances and decreased myocardial contractility were less frequently observed, along with other cardiac and hemodynamic changes. Cardiac and hemodynamic changes were more commonly observed in patients with chondromanubrial prominence.

Epidemiology

United States
Pectus excavatum is more common than the carinatum deformity. The overall prevalence of pectus carinatum is estimated at 0.06%. Fonkalsrud (2008) reported that at least 25% patients have a positive family history of chest wall deformity. Pectus carinatum can also be seen in association with Marfan syndrome and congenital heart disease.

International
The percentage of chest wall deformities represented by pectus carinatum are greater in reports from Brazil and Argentina.

Mortality/Morbidity
Psychological and cosmetic concerns are the most prominent reasons for initial consultation. However, Fonkalsrud (2008) reported that surgical repair is rarely performed only for cosmetic reasons. Morbidity in later years includes cardiac and hemodynamic changes.

Race
- The condition is more frequent in whites and is uncommon in blacks and Asians.

Sex
- Males are affected 4 times more frequently than females. Because this deformity may occur either in isolation or as part of a syndrome, identifying a single aetiology for the male predominance is difficult.

Age
- Although pectus carinatum has been described at birth, it is most frequently identified in mid childhood. The deformity often worsens during the adolescent growth spurt.

Clinical Presentation
Parents or the patient may report that pectus carinatum has been present since birth or early childhood, but most children present at age 11-15 years. The degree of deformity may worsen during adolescence, and most patients are asymptomatic. Once adult growth has occurred, the severity of the deformity generally remains stable.

Symptomatic patients report exertional dyspnoea and tachypnoea as well as decreased endurance. In one series, asthmatic symptoms were reported by 22% of patients. Musculoskeletal chest pain and tenderness when lying in the prone position can also occur. Patients may be affected by low self-esteem, poor body image, and decreased mental quality of life.

Physical
Two main types of pectus carinatum deformities have been described: chondrogladiolar and chondromanubrial. The most common is the chondrogladiolar form, in which there is a symmetric protrusion of the sternum and costal cartilages.
Some authors think that a lateral category should also be included. The image below shows an example of a child with a lateral deformity.

In addition to the descriptive findings of anterior chest wall prominence, poor chest wall expansion with inspiration may be observed.

**Causes**

Aetiology has not been established; however, the increased incidence of positive family history and associated anomalies has suggested an abnormality in connective tissue development. Pectus carinatum deformities are associated with overgrowth of the rib cage during development of the chest wall.

**Diagnostic Considerations**

Diagnosis of pectus carinatum is clinical and is based on descriptive findings identified during the physical inspection of the chest. It may occur as an isolated anomaly, in association with congenital heart disease, or with another skeletal anomaly (20% scoliosis). Mixed deformities can be observed in Poland syndrome. Approximately 25% of patients have a positive family history of chest wall deformity. Less frequently, pectus carinatum has been associated with Morquio syndrome, hyperlordosis, and kyphosis.

**Imaging Studies**

Radiographic imaging should include 2 view chest radiographs: posteroanterior and lateral images. A chest radiograph of a patient with pectus carinatum is shown in the image below.

Additional imaging with either a chest CT scan or MRI may also be helpful. CT scanning of the chest in an individual with pectus carinatum (see the image below) reveals an increased anteroposterior chest wall diameter.

The Haller method may be used to determine severity index, as follows: width of the chest divided by distance between the sternum and spine at the same level; this may help to predict those individuals who will benefit from surgical intervention.

**Other Tests**

In patients with pectus carinatum, pulmonary function studies may be tailored to address concerns about clinical symptoms and the appearance of the chest wall upon examination.
Data on pulmonary and exercise physiology in patients with pectus carinatum deformities are limited. However, children with barrel chests usually have obstructive ventilatory defects. This underscores the importance of performing complete pulmonary function testing, including prebronchodilator and postbronchodilator spirometry, lung volumes, and diffusion capacity. Exercise testing may complement these studies.

In 1982, Castile described one patient who reported exercise intolerance in his series of symptomatic pectus deformities. His pulmonary function studies revealed flow rates and lung volumes within the reference range. Derveaux's 1989 series also reported a patient with no significant respiratory compromise at the time of his study.

Progressive exercise studies may also be helpful in evaluating the exercise-related symptoms and exertional tolerance.

Electrocardiography and echocardiography may be considered if congenital heart disease is suspected. Iakovlev's study reported 70 patients with pectus carinatum deformity. Of these, 97% had echocardiographically documented mitral valve prolapse. Hemodynamic and cardiodynamic changes were also observed in some patients, as well as decreased myocardial contractility. These abnormalities were more frequently observed in the patients with pigeon breast. Scoliosis series may be considered if clinical features are suggestive of this diagnosis. Chromosomal analysis and metabolic testing may also be considered if other dysmorphic signs are identified.

**Medical Care**

Most motivated patients with pectus carinatum, especially those younger than 18 years with malleable chest walls, benefit from orthotic bracing, and this is generally the first line of therapy. Success rates of 65-80% and long-term outcomes with orthotic bracing alone are encouraging.

A study by Lee et al (2012) describes the preliminary results of 98 children treated using the Calgary Protocol, which involves a self-adjustable, low-profile bracing system used in 2 phases. The first phase involves 24 h/d bracing until correction is achieved. The second phase is a maintenance phase during which the brace is worn only at night until axial growth is complete. Twenty-three children completed treatment with good patient satisfaction and improved appearance, suggesting that when used in this fashion, it is an effective treatment for pectus carinatum. Therapy failed in 42 children, owing to either noncompliance or they were lost to follow-up. Two required surgical intervention after bracing failed to correct the problem. Additional studies and follow-up of the children still on protocol is important.

For older patients with more rigid chest walls, bracing may not be effective and surgery may be the initial consideration. Casting followed by bracing or bracing alone eliminates the risks of surgery and anaesthesia and does not preclude surgery if unsuccessful.

**Surgical Care**

Endoscopic resection of costal cartilage with a sternal osteotomy. Because many corrections are performed for cosmetic reasons, decreasing the size of incisions is important. In 1997, Kobayashi reported 2 patients in whom the pectus carinatum deformity was corrected with limited incisions using an endoscopic approach. They suggest that this approach is better
indicated in preschool-aged children because of their skin quality and tone, as well because of the increased ease of costal dissection compared with adult patients.

In 2008, Fonkalsrud reported a series of 260 patients who underwent surgical correction of pectus carinatum deformities over a period of 37 years. He concluded that, over time, the trend towards less extensive open techniques has resulted in "low morbidity, mild pain, short hospital stay and very good physiologic and cosmetic results." His study included both paediatric and adult patients.

**Consultations**

Pectus carinatum has been associated with congenital heart disease. In these patients, and in those with suspected or identified cardiac pathology, preoperative cardiology evaluation is recommended. Exercise testing may be performed in consultation with either a cardiologist or a pulmonologist. Symptomatic patients with exertional dyspnoea, tachypnoea, or decreased endurance, as well as those with asthma symptoms, benefit from a pulmonology evaluation. Individuals with pectus carinatum who have significant concerns about their body image or low self-esteem can benefit from psychological counselling.

**Activity**

Symptomatic patients may report decreased exercise tolerance and exertional dyspnoea, which may limit activity. Fonkalsrud's series (2008) reported improvement in exertional symptoms and endurance in all symptomatic patients within 3-6 months of surgical repair.

Fonkalsrud's recommendations for postoperative activity include the following.

- Use incentive spirometer and encourage periodic deep breaths.
- Limit twisting movements of the chest for at least 4 months postoperatively.
- Avoid rapid elevation of the arms overhead for at least 4 months postoperatively.
- Encourage lower extremity exercise (may begin within first 2 wk after surgery).
- Light weights may be used to strengthen biceps and deltoids; the use of chest and abdominal muscles may be increased later (after 3-4 wk).
- Gym classes are not indicated for 5 months after surgery in school-aged children.
- Long-term recommendations include stretching exercises that involve pulling the shoulder blades posteriorly to improve posture.
- Long-term activity recommendations include stretching.

**Complications**

Complications vary according to treatment selection. Ill-fitting braces can be associated with skin irritation and skin breakdown. Shamberger reported a 3.9% complication rate with open surgical repair. Complications include pneumothorax (2.6%), wound infection (0.7%), atelectasis (0.7%), and local tissue necrosis (0.7%). The mean postoperative stay was 5.8 days. Fonkalsrud (2008) reported shorter hospital stays (mean, 2.6 d), mild postoperative pain, and low complication rate with limited resection and immediate chest stabilization.

**Prognosis**

In prepubertal children with pectus carinatum who are compliant with bracing, success rates are excellent (up to 80%).
Excellent results (97.4%) have been reported by Fonkalsrud (2008) in patients who underwent surgical correction using a very limited resection of deformed cartilage and immediate chest stabilization. In addition, he reported less postoperative pain, shorter hospital stays, lower complication rate, and decreased cost. Furthermore, he reported satisfactory cosmetic results with the less extensive repair, as well as a high rate of improvement in exertional symptoms compared with more extensive open surgical procedures. Recurrences are rare.

Responses to quality-of-life questionnaires in patients who had undergone minimally invasive repair of their pectus deformity supported a positive impact on psychosocial function.

**Osteogenesis Imperfecta**

Osteogenesis imperfecta (OI) is a disorder of bone fragility chiefly caused by mutations in the \( \text{COL1A1} \) and \( \text{COL1A2} \) genes that encode type I procollagen. Four types of osteogenesis imperfecta were originally described by Sillence in 1979 and are now used broadly as the Sillence Criteria. The Nosology and Classification of Genetic Skeletal Disorders provides similar categorization in the 2010 revision. Precise typing is often difficult, and depends in large degree on the experience of the clinician. Severity ranges from mild forms to lethal forms in the perinatal period. Additional genes have been discovered in which mutations can also cause brittle bones. These are typically clinically indistinguishable and are considered by most to be subtypes of osteogenesis imperfecta. Examples of common radiologic findings of osteogenesis imperfecta are shown in the images below.
Pathophysiology

The most widely used classification of osteogenesis imperfecta published by Sillence et al in 1979 does not include additional forms of the disorder which have been discovered with improvements in molecular diagnostics. Rather, these forms of osteogenesis imperfecta are caused by genes which interact with collagen I or in the complex relationship between formation and remodelling of bone. They are molecularly distinct from osteogenesis imperfecta caused by COL1A1 or COL1A2 but form an example of locus heterogeneity. Forlino and Marini in 2015 offer an alternate way of understanding the genetics of osteogenesis imperfecta by sorting into five functional categories as follows:

- **Group A:** Primary defects in collagen structure or function (COL1A1, COL1A2, BMP1)
- **Group B:** Collagen modification defects (CRTAP, LEPRE1, PPIB, TMEM38B)
- **Group C:** Collagen folding and cross-linking defects (SERPINH1, FKBP10, PLOD2)
- **Group D:** Ossification or mineralization defects (IFITM5, SERPINF1)
- **Group E:** Osteoblast development defects with collagen insufficiency (WNT1, CREB3L1, SP7)

This classification system has not been integrated into widespread use but offers significant streamlining of categories into intellectually satisfying divisions.

**Osteogenesis Imperfecta with Calcification of the Interosseous Membranes** (Type V)

Patients with this form of osteogenesis imperfecta generally have moderate severity disease but frequently develop hyperplastic calluses in long bones after having a fracture or orthopaedic surgery that involves osteotomies. The size and shape of the callus may remain stable for many years after a rapid growth period, but in some cases slowly involutes. Patients also frequently develop calcification of the forearm interosseous membrane and dislocation of the radial head, which results in difficulties with supination and pronation. They may also have subphyseal metaphyseal radiodensity seen on radiography. This condition is caused by heterozygous mutations in the IFITM5 gene, and inheritance is autosomal dominant.

Histology of bone showed that the lamellae are arranged in an irregular fashion and in some cases appeared mesh-like, as opposed to the typical parallel arrangement in patients with osteogenesis imperfecta.
**Osteogenesis imperfecta with congenital joint contractures,** Types 1 and 2 (Bruck syndrome)

Patients with Bruck syndrome have congenital brittle bones prone to fracture, as well as congenital joint contractures and pterygia. They also have short stature, severe limb deformity, wormian bones, and progressive scoliosis which can be severe. Patients have generally been described with normal hearing, no dentinogenesis imperfecta, and white sclerae. Two forms of Bruck syndrome have been delineated with molecular testing, but appear to be clinically indistinguishable. Bruck syndrome 1 is caused by homozygous mutation in the *FKBP10* gene, while Bruck syndrome 2 is caused by homozygous mutation in the *PLOD2* gene. Both disorders are inherited in an autosomal recessive manner. It has been suggested that the defect underlying Bruck syndrome is a deficiency of bone-specific telopeptide lysyl hydroxylase, which results in aberrant bone collagen crosslinking.

**Epidemiology**

**Frequency**

**United States**
The prevalence of OI is estimated to be 1 per 15,000 live births; however, the mild form is underdiagnosed, and the actual prevalence may be higher.

**International**
Prevalence appears to be similar worldwide, although there may be an increased risk of recessive forms of osteogenesis imperfecta in populations with high degree of consanguinity.

**Race** - No differences based on race are reported.

**Sex** - No differences based on sex are reported.

**Age** - The age when symptoms (i.e., fractures) begin widely varies. Patients with mild forms may not have fractures until adulthood, or they may present with fractures in infancy. Patients with severe cases typically present with fractures in utero.

**History**
Patients sometimes have a family history of osteogenesis imperfecta (OI), but most cases are due to new mutations.

- Patients most commonly present with fractures after minor trauma.
- In severe cases, prenatal screening ultrasonography performed during the second trimester may show bowing of long bones, fractures, limb shortening, and decreased skull echogenicity. Lethal osteogenesis imperfecta cannot be diagnosed with certainty in utero.
- Patients may bruise easily.
- Patients may have repeated fractures after mild trauma. However, these fractures heal readily.
- Hearing loss is a variable feature of osteogenesis imperfecta. About 50% of patients with type I osteogenesis imperfecta have hearing loss by age 40 years.

**Physical**
Physical examination can vary depending on the severity. Degrees of severity may vary to some extent among different affected members of the same family.

**Type I - Mild, nondeforming**

See the list below:

- Patients have no long-bone deformity.
The sclera can be blue or white. Blue sclera may also occur in other disorders, such as progeria, cleidocranial dysplasia, Menkes syndrome, cutis laxa, Cheney syndrome, and pyknodysostosis.

Dentinogenesis imperfecta may be present.

Over a lifetime, numbers of fractures can range from 1-60 or more.

Height is usually normal in individuals with mild forms of osteogenesis imperfecta, but may be less than unaffected members of their family.

People with osteogenesis imperfecta have a high tolerance for pain. Old fractures may be discovered in infants only after radiographs are obtained for other reasons other than an assessment of osteogenesis imperfecta, and they can occur without any signs of pain.

Exercise tolerance and muscle strength are frequently reduced in patients with osteogenesis imperfecta, even in the mild forms.

Fractures are most common during infancy but may occur at any age.

Other possible findings include kyphoscoliosis, hearing loss, premature arcus senilis, and easy bruising.

Type II – Perinatal lethal

See the list below:

Some providers who treat large numbers of patients with osteogenesis imperfecta suggest that the diagnosis of Type II OI be made in retrospect for patients who do not survive the perinatal period, and that even patients with very severe forms of OI who nonetheless are long term survivors be classified as Type III.

Blue sclera may be present.

Patients may have a small nose, micrognathia, or both.

All patients have in utero fractures, which may involve the skull, long bones, and/or vertebrae.

The ribs are beaded, and the long bones are severely deformed.

Causes of death include extreme fragility of the ribs, pulmonary hypoplasia, and malformations or haemorrhages of the CNS.

Type III – Severe, progressively deforming

See the list below:

Patients may have joint hyperlaxity, muscle weakness, chronic unremitting bone pain, and skull deformities (e.g., posterior flattening) due to bone fragility during infancy.

Deformities of upper limbs may compromise function and mobility.

The presence of dentinogenesis imperfecta is independent of the severity of the osteogenesis imperfecta.

The sclera have variable hues.

In utero fractures are common.

Limb shortening and progressive deformities can occur.

Patients have a triangular face with frontal and temporal bossing. Malocclusion is common.

Basilar invagination is an uncommon but potentially fatal occurrence in osteogenesis imperfecta.

Vertigo is common in patients with severe osteogenesis imperfecta.
• Hypercalciuria may be present in about 36% of patients with osteogenesis imperfecta, and adults may be at higher risk of renal calculi.
• Respiratory complications secondary to kyphoscoliosis are common in individuals with severe osteogenesis imperfecta.
• Constipation and hernias are also common in people with osteogenesis imperfecta.

Type IV - Common variable

See the list below:

• This type of osteogenesis imperfecta is not as clearly defined as other types.
• Height is extremely variable with some patients having near-normal height and others having significantly short stature.
• Dentinogenesis imperfecta may be present. Some have suggested that this sign can be used to divide type IV osteogenesis imperfecta into subtypes a and b.
• Fractures usually begin in infancy, but in utero fractures may occur. The long bones are usually bowed, although typically not as severe as patients with type III.

Causes

Osteogenesis imperfecta is an inherited disorder. In Types I-V osteogenesis imperfecta, the mode of inheritance is autosomal dominant and often involves a new dominant mutation. This accounts for between 90-95% of the total disease burden. The remaining 5-10% of patients show an autosomal recessive inheritance pattern, with both parents generally being asymptomatic carriers.

Some have proposed possible germ-cell mosaicism as an explanation for cases occurring in families with healthy parents that have more than one child with osteogenesis imperfecta. In some cases, somatic mosaicism has been noted in parents who have had multiple children with the same dominant form of osteogenesis imperfecta.

Diagnostic Considerations

These include the following:

• Nonaccidental trauma
• Campomelic dysplasia
• Achondrogenesis type I
• Infantile/Perinatal/Juvenile hypophosphatasia
• Steroid-induced osteoporosis
• Idiopathic juvenile osteoporosis
• Osteoporosis of disuse

Differential Diagnoses

• Physical Child Abuse

Laboratory Studies

Results from routine laboratory studies in patients with osteogenesis imperfecta (OI) are usually within reference ranges and they are useful in ruling out other metabolic bone diseases such as hypophosphatasia or inherited forms of rickets.

Collagen synthesis analysis is performed by culturing dermal fibroblasts obtained during skin biopsy. The occurrence of false-negative results is not clear, although the rate may be about
15%. As molecular diagnostics have become more widespread, less expensive, and covered to an increasing extent by third-party payers in the United States, this testing has fallen out of fashion.

Prenatal DNA mutation analysis can be performed in pregnancies with risk of osteogenesis imperfecta to analyse uncultured chorionic villus cells. Samples are obtained during chorionic villus sampling performed under ultrasonographic guidance when a mutation in another member of the family is already known.

Bone mineral density, as measured with dual-energy radiographic absorptiometry (DRA), is generally low in children and adults with osteogenesis imperfecta. However, there is wide variation in the bone density of patients with OI. Still, normal bone density in a patient whom osteogenesis imperfecta is being evaluated should prompt consideration of alternate diagnoses.

**Imaging Studies**

Obtain a radiographic skeletal survey after birth.

- In mild (type I) osteogenesis imperfecta, images may reveal thinning of the long bones with thin cortices. Several wormian bones may be present. No deformity of long bones is observed.
- In extremely severe (type II) osteogenesis imperfecta, the survey may reveal beaded ribs, broad bones, and numerous fractures with deformities of the long bones. Platyspondylia may also be revealed.
- In moderate and severe (types III and IV) osteogenesis imperfecta, imaging may reveal cystic metaphyses, or a popcorn appearance of the growth cartilage. Normal or broad bones are revealed early, with thin bones revealed later. Fractures may cause deformities of the long bones. Old rib fractures may be present. Vertebral fractures are common.

Prenatal ultrasonography can be used to detect limb-length abnormalities at 15-18 weeks of gestation.

- Mild forms may result in normal sonogram findings.
- Features include super-visualization of intracranial contents caused by decreased mineralization of calvaria (also calvarial compressibility), bowing or fracture of the long bones, decreased bone length (especially of the femur), and multiple rib fractures.

**Histologic Findings**

See the list below:

- The width of biopsy cores, the width of the cortex, and the volume of cancellous bone are decreased in all types of osteogenesis imperfecta. The number and thickness of trabeculae are reduced.
- Samples may show evidence of defects in modelling of external bone in terms of the size and shape, the production of secondary trabeculae by endochondral ossification, and the thickening of secondary trabeculae by remodelling. Therefore, osteogenesis imperfecta might be regarded as a disease of the osteoblast.
- Bone formation is quantitatively decreased, but the quality of the bone material is probably most important in the pathogenesis of the disease.
Medical Care

Because osteogenesis imperfecta (OI) is a genetic condition, it has no cure.

- Cyclic administration of intravenous pamidronate reduces the incidence of fracture and increases bone mineral density, while reducing pain and increasing energy levels. Doses vary from 4.5-9 mg/kg/y, depending on the protocol used.
- Current evidence does not strongly support the use of oral bisphosphonates in patients with osteogenesis imperfecta, although they are sometimes used. Risedronate may have some effect in reducing fractures in patients with osteogenesis imperfecta.
- A preclinical study demonstrated that RANKL inhibition improves density and some geometric and biomechanical properties of the oim/oim mouse bone but does not decrease fracture incidence when compared with placebo. Research into the RANK inhibitor denosumab in patients with type VI OI has produced positive results, although only 2 years of follow-up have been published.
- Nutritional evaluation and intervention are paramount to ensure appropriate intake of calcium and vitamin D. Caloric management is important, particularly in adolescents and adults with severe forms of osteogenesis imperfecta.
- In utero transplantation of adult bone marrow has been shown to decrease perinatal lethality in a murine model of osteogenesis imperfecta.

Surgical Care

Orthopaedic surgery is one of the pillars of treatment for patients with osteogenesis imperfecta. Surgical interventions include intramedullary rod placement, surgery to manage basilar impression, and correction of scoliosis.

Intramedullary rod placement

- In patients with bowed long bones, intramedullary rod placement may improve weight bearing and, thus, enable the child to walk at an earlier age than he or she might otherwise. Use of the extensible Fassier-Duval rod has been very helpful in improving bowing and mobility in patients with osteogenesis imperfecta.
- In children appropriately treated with bisphosphonates, the percutaneous technique of multiple osteotomy with intramedullary fixation is safe and effective.
- An experienced team can perform as many as 4 rod procedures in the long bones of the lower extremities in one surgical session.
- Fractures heal normally in about 85% of patients with osteogenesis imperfecta.
- Postoperative immobilization is significantly shortened with this technique. Prolonged immobilization after a fracture must be avoided.

Surgery for basilar impression: This procedure is reserved for cases with neurologic deficiencies, especially those caused by compression of brainstem and high cervical cord. A team of orthopaedic surgeons and neurosurgeons is required.

Correction of scoliosis: Correction of scoliosis may be difficult because of bone fragility. Spinal fusion surgery can be beneficial in patients with severe disease.

Neurofibromatosis Type 1

The clinical features of neurofibromatosis type 1 (NF1) or peripheral neurofibromatosis, the most common form of the disease, were reported in several family members by German
pathologist Virchow in 1847, but it was his student von Recklinghausen who 35 years later described the histologic features of the syndrome that often bears his eponym.

The orthopaedic manifestations and, especially, the complications after treatment of NF1 are common and have a prominent place in the orthopaedic literature. The intent of this article is to identify the complications most commonly associated with the orthopaedic manifestations of neurofibromatosis and to present strategies for their management.

Skeletal complications of NF1 can be categorized as generalized or focal manifestations.

Generalized skeletal abnormalities include osteoporosis/osteopenia, osteomalacia, shortness of stature, and macrocephaly. The focal orthopaedic complications of neurofibromatosis, which usually appear early, include spinal deformities such as scoliosis and kyphoscoliosis, congenital bowing and pseudoarthrosis of the tibia and the forearm, chest wall deformities, overgrowth phenomenon of the extremity, and soft-tissue tumours. (See the images below.) Focal abnormalities of the skeleton are less common than generalized abnormalities are but may cause significant morbidity.

Neurofibromatosis type 2 (NF2) or central neurofibromatosis is associated with bilateral vestibular schwannomas and multiple spinal schwannomas. NF1 and NF2 are genetically distinct disorders despite the similarity of their names.

Segmental neurofibromatosis is characterized by features of NF1 involving a single body segment. Typically, only a single segment of the body (such as left upper extremity) is affected with café-au-lait spots and freckling, and lesions usually do not cross the body midline.

Early neurofibromatosis literature recognized that a mild form of NF1 existed, consisting primarily of familial café-au-lait spots. In recent years, multiple families with such mild involvement have now been found to have mutations in the SPRED1 gene. This condition, now called Legius syndrome, can present with multiple café-au-lait spots, freckling, macrocephaly, and mild learning disabilities, but does not present with any of the benign or malignant tumours associated with NF1.

Schwannomatosis is a distinct form of neurofibromatosis that typically involves multiple schwannomas throughout the body, but without the vestibular schwannomas typical of NF2. It is most commonly a disease of adulthood that consists of multiple deep painful peripheral nerve sheath tumours, which may occur in a generalized form or in a segmental distribution.
NF2, segmental neurofibromatosis, Legius syndrome, and schwannomatosis do not appear to have any bone involvement or other orthopaedic manifestations. As a result, they are not discussed in this article.

Spinal Complications

Spinal complications are the most common orthopaedic manifestation of NF1. Between 10% and 33% of children with NF1 have spinal deformity. Deformities include dystrophic and non-dystrophic changes. The radiologic appearance of the dystrophic changes includes the following features:

- Scalloping of the vertebral body margins (3 mm in thoracic vertebrae, 4 mm in lumbar vertebrae, or both)
- Severe rotation of the apical vertebra
- Widening of the spinal canal
- Enlargement of the neural foramina
- Defective pedicles
- A paraspinal mass
- Spindling of the transverse process
- Rib pencilling (i.e., the rib being smaller in diameter than the second rib) - The ribs may resemble twisted ribbons
- Presence of dural ectasia

More than three of these dystrophic features are considered diagnostic of dystrophic scoliosis. In some cases, these changes are the result of intraspinal pathology, such as tumours, meningoceles, or dural ectasia. However, the changes may also occur in persons with entirely normal intraspinal contents. In these persons, primary bone dysplasia accounts for the dystrophic changes.

Spinal changes in individuals with NF1 are usually divided into cervical, thoracic, lumbosacral, and spinal canal pathologies.

Cervical spine changes and associated complications

Features of the cervical spine in patients with NF1 have not received enough attention in the literature. Cervical abnormalities occur much more frequently when a scoliosis or kyphoscoliosis is present in the thoracolumbar region, in which case the examiner’s attention is focused on the more obvious deformity.

The manifestations of NF1 can be observed as dystrophic changes in the vertebral body, or they can be due to pathologic alignment. The most common abnormality observed is a severe cervical kyphosis, which in itself is highly suggestive of the disorder.

In a study by Yong-Hing et al, 17 patients with NF1 were found to have cervical abnormalities. Of these, seven were asymptomatic, and the rest had either limited motion or pain in the neck. Four patients had neurologic deficits, which were probably attributed to cervical instability. Four of the 17 patients required fusion of the cervical spine. Curtis described eight patients with paraplegia and NF1. Paraplegia in four of these patients was due to cervical spine instability or intraspinal pathology in the cervical spine.

Attention also should be paid to the C1-2 region. Isu et al. described three patients with NF1 who had C1-2 dislocation with neurologic deficit, and all improved after decompression or fusion. No bony changes in the C1-2 relationship were observed on flexion-extension views in any of these patients. Most of the problems in the cervical spine in this study occurred
after excision of the tumours, which included resection of the laminae and posterior elements. Postoperatively, the spine is unstable and tends to develop progressive kyphosis.

All patients with NF1 who undergo surgery, who require endotracheal anaesthesia, who undergo halo traction, who complain of neck pain, or who present with neck tumours should undergo a cervical radiographic series. If subluxation is suspected, computed tomography (CT) scans, magnetic resonance imaging (MRI) scans, or both are appropriate. Other reasons for obtaining cervical spinal radiographs in a patient with NF1 include the evaluation of torticollis and dysphagia (symptoms indicating intra- or extraspinal neurofibromas).

There is significant progressive kyphosis following occipitocervical decompression in young children. Deformity progression is silent and may not be noted. Nothing less than biannual imaging follow-up is mandatory. Any patient with any evidence of instability after an operative procedure on the spine that requires laminectomy should undergo spinal fusion.

**Scoliosis**

Scoliosis is the most common osseous defect associated with NF1. It may vary in severity from mild and non-progressive to severe curvatures. The cause of this spinal deformity is unknown, but some have suggested that it is secondary to endocrine disturbances, mesodermal dysplasia, and osteomalacia (a localized neurofibromatous tumour eroding and infiltrating bone).

In a general orthopaedic clinic, 2% of patients with scoliosis have neurofibromatosis, whereas in a neurofibromatosis clinic, 10-20% of patients have some disorder of the spine. All preadolescent children with neurofibromatosis should be evaluated with scoliosis screening, or the Adams bend test, to exclude a spinal deformity, which usually occurs earlier in children with neurofibromatosis.

Two primary types of scoliosis are observed in persons with neurofibromatosis: dystrophic and non-dystrophic. Dystrophic scoliosis is the short-segmented, focal, sharply angulated type that includes fewer than six spinal segments. It has a tendency to progress to a severe deformity. Dystrophic curves may be associated with kyphosis and have a higher incidence of neurologic injury.

The second type of curvature, non-dystrophic scoliosis, is similar to the idiopathic curvature observed in adolescents. This form usually involves eight to 10 spinal segments. The deformity is most often convex to the right; however, this is not consistent. Compared to dystrophic curves, non-dystrophic curves tend to present in older children with less angulation and rotation of the deformity.

Patients more likely to develop progressive scoliosis are children younger than 7 years who have thoracic lordosis (sagittal plane angle <20°) and paravertebral tumours. Curves that acquire either three or more pencilled ribs or a combination of any three dystrophic features as described above (modulation theory), will almost certainly progress. Durrani et al defined modulation as a process by which a non-dystrophic curve acquires the features of a dystrophic curve and behaves as a dystrophic curve. They reported that modulation occurred in about 65% of their patients.

Subsequent MRI studies questioned the theory of modulation. Patients with radiographically labelled non-dystrophic curves were found to have significant dysplastic changes on MRI. Given that MRI has higher sensitivity for identifying dystrophic features than x-rays do, it is strongly recommended to characterize the curve as dystrophic or not on the basis of a combination of MRI and x-ray findings.
Meningoceles, pseudomeningoceles, dural ectasia, and dumbbell lesions are all related to the presence of neurofibroma or abnormal pressure phenomena in and around the spinal canal neuraxis. High-volume or contrast MRI should be used in the investigation of all dystrophic curves prior to treatment. Occasionally, these intraspinal elements may directly compromise the cord when instrumentation and stabilization are attempted, or they may cause erosive changes in the bone, preventing primary fusion.

The cervical spine should be evaluated with the initial scoliosis investigation. Evidence of dystrophic changes may be present on a true lateral view. Progressive cervical kyphosis is usually apparent after excision of the posterior elements. The patient presents with a neck deformity after anterior and posterior excision of a neck mass. If any suspicious area is noted on plain radiographs, right and left oblique views should be obtained to look for widening of the neuroforamina. These may represent dumbbell lesions (i.e., widening of the neuroforamina caused by dural ectasia or the exit of a neurofibroma from the spinal canal).

**Kyphosis**

Kyphosis observed in individuals with NF1 is distinguished by acute anteroposterior angulation. Vertebral bodies may be deformed so severely that they are confused with congenital deformities. It may occur by gradual scoliotic rotation and progression or it can be found early in the disease with an abrupt angular kyphotic curve. Severe kyphosis is the most common cause of neurologic deficits in NF1.

**Lordoscoliosis**

Lordoscoliosis has not been as frequently reported in patients with NF1 as kyphoscoliosis has. However, lordosis of the thoracic spine predisposes to significant respiratory compromise and mitral valve prolapse.

**Spondylolisthesis**

Spondylolisthesis is a rare disorder that is most often associated with a pathologic luxation of the vertebra because of erosions of the pedicles or pars from foraminal neurofibroma or dural ectasia.

**Tibial Dysplasia**

Tibial bowing occurs in 1 per 140,000 live births. The bowing associated with NF1 is always anterolateral. The deformity may appear before other protein manifestations, such as café-au-lait spots. It is usually evident within the first year of life, with a fracture not uncommonly occurring by the time the child is aged 2-2.5 years. Conversely, posteromedial congenital bowing, or kyphoscoliosis tibia, is a benign condition.

Tibial bowing associated with skin dimpling, ring constrictions, and foot deformities is rarely associated with NF1. The management of this anterolateral bowing deformity is most frustrating. Unlike scoliosis, treatment of congenital pseudoarthrosis of the tibia does not appear to be more successful when it is initiated early.

There are two basic types of bowing: nondysplastic and dysplastic. Nondysplastic (type I) bowing is defined as follows:

- Anterolateral bowing with increased bony density
- Sclerosis of the medullary canal
- Possibility of this type converting to dysplastic after fracture or osteotomy to correct the angulation

Dysplastic (type II) bowing is defined as follows:

- Anterolateral bowing with a failure of tubulation
• Anterolateral bowing with cystic prefracture or canal enlargement from previous fracture
• Frank pseudarthrosis and atrophy with “sucked candy” narrowing of the ends of the two fragments

Scoliosis treatment

The treatment of non-dystrophic curvatures is very similar to that of idiopathic scoliosis. Curves of less than 25° should be observed. Curves between 25° and 40° can occasionally be treated successfully with bracing. Once beyond 40°, surgery by posterior spinal fusion is usually indicated.

Dystrophic curvatures of less than 20° should be observed for progression at 6-month intervals. For persons with curvature greater than 20-40° of angulation, a posterior spinal fusion with some form of segmental spinal instrumentation is recommended. [14] Curves greater than 50° should be treated with anterior and posterior fusion.

Oblique radiographs are obtained every 6 months to exclude pseudarthrosis. Brace treatment has not been consistently effective.

For the very young child, early fusion causes minimal stunting of growth. Furthermore, the dystrophic segments have very limited growth potential to begin with. In these young children, non-dystrophic curvatures of less than 20° should be observed, those of 20-35° should be braced, and those of 35° or greater should be stabilized by means of anterior and posterior fusion with segmental spinal instrumentation.

Growing rods have been used in very young children with NF1 to obtain correction without definitive fusion and to lengthen or “grow the spine” every 6 months, but with varying success and a high rate of complications. The MAGEC Spinal Bracing and Distraction System (NuVasive, Aliso Viejo, CA), which does not require operative intervention for lengthening, may be beneficial in managing the skeletally immature patient, but there is no current experience with this approach.

Kyphosis

Bracing is recommended for patients with kyphosis of less than 50°. In patients with curvatures greater than 50° that fail to correct on cross-table lateral x-rays taken hyperextended over a bolster, anterior surgery (intervertebral discectomy, rib strut grafting, and bone chip grafting) is required, followed by posterior segmental instrumentation. Once a curvature exceeds 70° and fails to correct as above, surgery is strongly indicated. Bracing is required after spinal surgery until there is evidence of bony fusion.

Because of the association of paraplegia with kyphosis, physicians have tended to perform laminectomy. Laminectomy alone for kyphotic cord compression is absolutely contraindicated. The offending neurofibromas are usually anterior, and decompression should be performed anteriorly. The removal of the posterior element predisposes the spine to instability by removing valuable bone stock required for fusion. Spinal fusion should always be performed after laminectomy.

Lordoscoliosis

Anterior release and intervertebral fusion followed by posterior instrumented fusion is considered as the most reliable surgical option to achieve correction of dystrophic lordoscoliosis. Sublaminar wires, pedicle screws, Universal Clamps (Zimmer Spine, Bordeaux, France), or rod-multiple hook constructs can be used.
Spondylolisthesis
Anterior and posterior stabilization is recommended for progressive deformity. Progression of spondylolistheses from grade II to grade III or the presence of pathologically elongated pedicles with or without pain are indications for surgery. Fusion may be delayed because of the forward traction effect of the vertebral bodies and the slow healing and remodelling of bone in NF. Postoperative immobilization is indicated until the fusion is absolutely solid. Dural ectasia may completely erode the pedicles in this condition and continue to do so after a successful spinal fusion.

Spinal Tumours
Primary spinal tumours fall into a distinct category because their timely diagnosis and the immediate institution of treatment have an enormous impact on the patient's overall prognosis and hope for a cure.

Generally, with spinal pathology, problems that arise are either chronic problems related to degenerative disease or deformity or acute manifestations of traumatic sequelae. When considering tumours of the spine, one must consider the different tissue types around the spinal column. The presence of neural tissue, meningeal tissue, bone, and cartilage makes any of these tissue types a possible nidus for neoplastic change. Also, metastatic lesions may spread to the spine from distant primary tumour sites by hematogenous or lymphatic routes.

Primary nonlymphoproliferative tumours of the spine are uncommon and make up fewer than 5% of bone neoplasms, accounting for fewer than 2.5-8.5 primary spine tumours per 100,000 people per year. Metastatic disease of the spine is much more common. Approximately 40-80% of patients who die of cancer have bony metastases at the time of death, with the spine being the most common metastatic skeletal location.

Neoplastic disease, however, can present with back pain that is indistinguishable from back pain resulting from more benign causes. Therefore, the physician caring for patients complaining of back pain is faced with the challenge of distinguishing benign causes from those that can be neurologically or systemically devastating and prescribing the appropriate treatment.

This distinction sometimes can be difficult to make because of the complicated architecture of the spine. The physician must consider differential diagnoses of degenerative processes, infections, muscular strains, neurologic impingements, and, finally, neoplastic processes. With thorough history taking, physical examination, and diagnostic imaging, the physician can acquire enough information to efficiently make the correct diagnosis.

Clinical Presentation
The most common clinical presentation associated with all spine tumours is back pain that causes the patient to seek medical attention. Back pain is the most frequent symptom for patients with either benign or malignant neoplasms of the spine. Neurologic deficits secondary to compression of the spinal cord or nerve roots also can be part of the presentation.

The degree of neurologic compromise can vary from slight weakness or an abnormal reflex to complete paraplegia, depending on the degree of encroachment. The loss of bowel or bladder continence can occur from neurologic compression or can be secondary to a local mass effect from a tumour in the sacrococcygeal region of the spine, as occurs in chordomas. Systemic or constitutional symptoms tend to be more common with malignant or metastatic disease than in benign lesions.
Laboratory studies

For these patients, workup should include a complete blood count and differential, a basic serum chemistry profile, erythrocyte sedimentation rate (ESR), or C-reactive protein (CRP) to help distinguish between neoplastic and infectious processes. Elevations in serum calcium or alkaline phosphatase also can provide evidence for neoplastic bone processes. Specific studies, such as serum electrophoresis or urine electrophoresis, also can be performed to evaluate the likelihood of multiple myeloma or plasmacytoma.

Imaging studies

Imaging studies for the workup of spine tumours include X-Ray, computed tomography (CT), magnetic resonance imaging (MRI), and technetium bone scanning.

The first-line imaging study should be plain radiography to evaluate the trabecular architecture of the spine. Anteroposterior (AP), lateral, and oblique views may be required. These studies should be evaluated with respect to both what the tumour is doing to the bone and, conversely, what the bone is doing to the tumour. The blastic or lytic nature of the lesion should be noted. The general location of the lesion within the bone, the integrity of the cortex, and the presence of fractures or soft-tissue masses are important findings (see the images below).

Figure 24 Spinal Tumours - Coned down view of haemangioma in thoracic spine

Figure 25 Spinal Tumours - Axial CT scan of haemangioma in lumbar vertebra
**Biopsy**

The ultimate way of making the diagnosis and ascertaining the specific tumour type is to perform a biopsy of the spine lesion after all radiographic studies have been completed. Biopsies can be performed with open technique or percutaneous image-guided technique. Percutaneous needle biopsies may not supply adequate tissue for the diagnosis of a primary tumour of the spine.

The basic principles of biopsy technique also apply to tumours of the spine. The surgeon performing the biopsy should take the most direct route to the tumour, with the least potential to contaminate adjacent compartments. The biopsy tract should be placed in line with the future incision site for surgical resection of the tumour, so that the biopsy tract can be excised with the specimen en bloc. (See the image below.)

![Figure 26 Spinal Tumours - photo of patient’s back at time of surgery](image)

Spinal tumours. Photograph of patient's back at time of surgery, exhibiting course of definitive incision to excise chondrosarcoma en bloc with previous biopsy tract included with resection.

Meticulous haemostasis must be obtained, and a drain must be placed to prevent hematoma formation, which can dissect the soft-tissue planes and contaminate adjacent compartments. The drain should exit the skin in line with the incision so that it, too, can be excised with the final specimen.

**Histologic findings**

The histologic findings vary according to the tumour types as described above. The following list revisits the primary tissue type associated with some of the tumours of the spine:

- Bone-producing tumours
- Cartilage-producing tumours
- Lymphoproliferative tumours
- Tumours of notochordal origin
- Round cell tumours

Bone-producing tumours of the spine include the following (see the images below):

- Osteoid osteoma - Benign and locally self-limited
- Osteoblastoma - Benign but locally expansile and aggressive
- Osteosarcoma - Malignant spindle cell lesion that produces osteoid

Cartilage-producing tumours of the spine include the following:

- Osteochondroma - Benign lesion with cartilaginous cap as described above
- Chondrosarcoma - Malignant cartilage producing tumours that histologically demonstrate round cellular stroma in a chondroid matrix (see the image below)
Lymphoproliferative tumours include the following:

- Multiple myeloma and plasmacytoma - Derived from plasma cell dyscrasias, which histologically appear as sheets of plasma cells
- Lymphoma - Associated with a large infiltrate of lymphoid cells

Chordoma is a tumour of notochordal origin that may be identified by the characteristic physaliferous cells (see the images below)

Ewing sarcoma is a malignant round cell tumour of childhood that is associated with large sheets of homogenous small, round, blue cells.

**Relevant Anatomy, Limiting Factors, and Staging**

The spine consists of 33 vertebrae that form the bony spinal column. The spinal column can be divided into the cervical, thoracic, lumbar, and sacrococcygeal regions. Although morphologically distinct, each vertebra in the subaxial cervical, thoracic, and lumbar spine has a complex architecture, consisting of a vertebral body, pedicles, laminae, and spinous and transverse processes.

The bony canal provides protection and support to the fragile spinal cord and nerve roots within the dural sac. The soft tissues surrounding the bony spine vary by location from the thick dorsal paraspinous musculature to the vital organs and vessels within the mediastinal, thoracic, peritoneal, and retroperitoneal spaces.

The relevant anatomy discussed above is frequently the limiting factor in determining contraindications for surgical excision of spine tumours. The morbidity of the tumour, the tumour’s malignant potential, and the patient’s overall prognosis must be compared to the morbidity and potential mortality of radical resection of a tumour near the spinal cord, the aorta, or the heart.

The degree of associated blood loss and the overall health of the patient also must be taken into consideration in considering a resection. If the patient is known to have metastatic or systemic tumour involvement, this may be a contraindication to radical resection of a paraspinous tumour, which may render the patient paralyzed.

Weinstein and McLain, and Boriani et al developed a descriptive staging system for spine tumours based on the principles of the Enneking staging system for primary bone tumours of the extremities. In this staging system, the transverse extension of the vertebral tumour is described with reference to 12 radiating zones (numbered 1-12 in clockwise order) and five concentric layers (A to E) from the paravertebral extraosseous compartments to the dural involvement. The longitudinal extent of the tumour is recorded according to the levels involved.

Based on an understanding of the biologic behaviour of the tumour, the oncologic staging aids the surgeon to decide what surgical margin provides the best chance for complete tumour resection and possible cure. This system is complex and sometimes difficult to apply clinically.

**Primary Benign Spinal Tumours**

The Enneking classification of benign lesions applies to benign spine tumours, as follows:

- Stage 1 - Latent
- Stage 2 - Active
- Stage 3 - Aggressive
Stage 1 lesions are usually asymptomatic and are discovered incidentally. Stage 2 lesions usually present with symptoms; most commonly, pain is in the area of the lesion. Stage 3 lesions are locally aggressive and can actually metastasize.

**Enostosis**

Also termed a bone island, enostosis is a mass of calcified medullary defects of lamellar compact bone with Haversian systems found within the cancellous portion of the bone. It occurs most frequently in the thoracic and lumbar spine, usually between T1 and T7 and between L1 and L2. Enostosis is one of the most common lesions to involve the spine.

Enostoses are usually stage 1 lesions and are discovered incidentally. Most remain stable, but some may slowly increase in size. Resnik et al determined the incidence of enostosis to be approximately 14% in cadavers.

Radiographically, enostoses are circular or oblong osteoblastic lesions with a spiculated margin, which gives it the appearance of thorny periphery. An abrupt transition from normal to the sclerotic bone is exhibited on the x-ray. Bone scan findings are usually normal, and magnetic resonance imaging (MRI) demonstrates low signal intensity with normal surrounding intensity.

Enostosis sometimes can be confused with osteoblastic metastatic disease. Enostosis can be differentiated by lack of activity on bone scan, by the normal appearance of adjacent bone, by its thorny margins, and by lack of a primary tumour for metastasis. If the enostosis exhibits an increase in diameter of greater than 25% in 6 months, a biopsy should be performed.

**Osteoid osteoma**

Osteoid osteomas usually present in children aged 10-20 years, with a male predominance. They involve the axial skeleton only 10% of the time. In the spine, 59% of osteoid osteomas are found in the lumbar region, 27% in the cervical region, 12% in the thoracic region, and 2% in the sacral region.

Osteoid osteomas are usually stage 2 lesions and are actively symptomatic. They can result in painful scoliosis, radicular pain, gait disturbances secondary to pain and splinting, and muscular atrophy. Symptoms usually are relieved or ameliorated by nonsteroidal anti-inflammatory drugs (NSAIDs) or salicylates. In the spine, osteoid osteomas occur 75% of the time in the posterior elements (pedicles, facets, or laminae). Osteoid osteomas occur 7% of the time in the vertebral body and 18% of the time in the transverse and spinous processes.

On plain x-ray, osteoid osteomas appear as a round or oval radiolucent nidus, with a surrounding rim of sclerotic bone. An area of central calcification may be present, but this classic appearance may be obscured by complex spinal architecture. Bone scan shows marked increased uptake by the nidus, and a double intensity pattern may exist. Computed tomography (CT) is the criterion standard for radiographic diagnosis. The nidus is a well-defined area of low attenuation with or without central calcification surrounded by an area of sclerosis. (See the image below.)
The nidus is usually smaller than 1.5-2.0 cm, composed of microscopic well-organized trabecular bone with vascular fibrous connective tissue stroma surrounded by reactive cortical bone.

Treatment is accomplished by resection of the nidus via an open surgical approach or by percutaneous CT-guided resection. Percutaneous radiofrequency ablation of the nidus has been performed with acceptable results.

**Osteoblastoma**

Histologically similar in appearance to osteoid osteoma, osteoblastoma is behaviourally very different. Demographically, it occurs in young patients in the second or third decade of life. A 2:1 male-to-female predominance exists. The lesion is distributed equally in the cervical, thoracic, and lumbar segments of the spine. The posterior elements are involved in 55% of cases, but the tumour can extend to the vertebral body in 42% of cases.

Patients typically complain of dull localized pain and paraesthesias, paraparesis and, if the tumour is large enough and encroaching on the spinal cord, paralysis.

Osteoblastomas are expansile lesions with multiple small calcifications and a peripheral scalloped and sclerotic rim. In more aggressive lesions, osseous expansion, bone destruction, infiltration of the surrounding tissue, and intermixed matrix calcification are present. Some 50% of osteoblastomas are radiolucent, and 20% are osteoblastic.

Marked radionucleotide uptake is exhibited on bone scan. CT demonstrates areas of mineralization, expansile bone remodelling, and sclerosis or a thin osseous shell at its margins. MRI is nonspecific but is the criterion standard to assess the effect of the tumour on the cord and surrounding tissues.
Osteoblastomas are typically larger than 2.0 cm in diameter with histologic features of interconnecting trabecular bone and fibrovascular stroma similar to, but not as well organized as, osteoid osteoma. They can have an aneurysmal bone cyst component in 10-15% of cases.

Wide local resection is the treatment of choice whenever possible. This sometimes is limited by the proximity of vital vessels or neural tissue in the spine. A 10-20% recurrence rate exists for conventional osteoblastomas. Aggressive osteoblastomas have a recurrence rate of approximately 50% if wide margins are not attained. These tumours are not radiosensitive.

**Aneurysmal bone cysts**

Aneurysmal bone cysts (ABCs) typically affect young patients, with 80% occurring in people younger than 20 years. The spine is involved 12-30% of the time. The thoracic spine is affected most commonly, followed by the lumbar and cervical spines. Sacral involvement is rare.

ABCs of the spine usually present as expansile areas of bone remodelling in the posterior elements. Extension into the vertebral body can occur 75% of the time. The lesion may have a thin outer periosteal rim of bone, and septations within the mass may be apparent. The mass may extend into adjacent vertebrae, discs, ribs, and paravertebral soft tissues.

The bone scan exhibits peripheral increased uptake with a central "cold area" creating a donut sign. If angiography is performed, the mass is found to be hypervascular 75% of the time. CT and MRI are used to confirm the cystic nature of the lesion as well as the tumour extension into surrounding tissues and the tumour’s relationship to the spinal canal. Single or multiple fluid/fluid levels sometimes can be visualized on MRI. MRI with gadolinium demonstrates enhancement of the periosteal rim and septations and not the cystic spaces.

ABCs are characteristically multiloculated blood-filled spaces that are not lined by endothelium. They are not vascular channels. Primary ABCs are believed to result from microtrauma to the bone with local circulatory disturbance. Other underlying neoplasms, such as giant cell tumours (GCTs), osteoblastomas, chondroblastomas, or osteosarcomas,
produce secondary ABCs. These other neoplasms produce venous obstruction and possible arteriovenous malformations and set the stage for ABC formation. Most ABCs are considered primary (65-95%).

Because of the locally aggressive behaviour of spinal ABCs, their treatment can be problematic. The severe morbidity that can be associated with complete resection is caused generally by danger to surrounding vascular or neural elements. ABCs can have a recurrence rate of 20-30% or higher, depending on the degree of resection. Preoperative embolization therapy and radiation may help shrink the tumour's size and decrease the amount of intraoperative blood loss associated with resection.

**Osteochondroma**
Osteochondromas make up 4% of all solitary spine tumours. They also are commonly referred to as exostosis. Spinal lesions are encountered in 7-9% of patients with multiple hereditary exostoses (MHE). Osteochondromas occur in patients aged 20-30 years. Patients with MHE tend to develop the osteochondroma at a younger age; they also tend to experience neurologic deficits and myelopathy more frequently (77% of the time) than the patient with solitary osteochondroma (34%). A male predominance exists.

Osteochondromas are more common in the cervical spine, especially at C2. The posterior elements usually are involved. The lesions are believed to arise secondary to trapping of the phyleal cartilage outside the growth plate during skeletal development.

Making the diagnosis of osteochondroma in the spine on plain radiography can be difficult unless the lesion is large and protruding posteriorly from a spinous process. In fact, 15% of patients with osteochondromas of the spine have normal appearing x-rays. CT is the study of choice to detect the exostosis and determine its relationship to the surrounding soft tissue and spinal canal.

T1-weighted MRI reveals a central area of high signal intensity, which represents yellow marrow. This area has intermediate intensity on T2-weighted images. The cortex of the exostosis has low signal intensity. The hyaline cartilage cap of the exostosis is best evaluated with MRI and appears with low signal intensity on T1 and high intensity on T2. The cartilage cap should be less than 2 cm in adults. Lesions with cartilage caps greater than 2 cm should be suspected of malignant transformation to chondrosarcoma.

Qualitatively, the bone composing an osteochondroma is normal. Abnormal bone growth occurs at and as a result of the cartilage cap. A continuity of the lesion with the marrow and cortex of the underlying bone is present. The exostosis may be sessile or pedunculated.

Complete surgical resection is usually curative. Clinical symptoms improve in 89% of patients following removal of the exostosis. Incomplete resection can lead to recurrence of the lesion.

**Giant cell tumour**
Giant cell tumours of the spine account for only 7% of all GCTs in the body. The spine is the fourth most common site for the occurrence of GCTs. Most spinal GCTs occur in the sacrum, followed by the thoracic, cervical, and lumbar regions. GCTs are more common in women and occur in the third to fifth decades of life. They can increase dramatically in size during pregnancy secondary to hormonal influences. Symptoms include pain with radicular pattern. With neurologic impingement, weakness and sensory deficits also can be manifest.

Spinal GCTs are usually radiolucent and expansile lesions. They do not exhibit active matrix production. When present in the sacrum, these lesions are large with destruction of the
sacral foraminal lines on plain x-rays. GCTs usually can involve both sides of the midline and can extend past the sacroiliac joints bilaterally. When present in sites proximal to the sacrum, they usually are found in the vertebral body.

The classic findings of GCT on technetium bone scan include diffuse radionuclide uptake with areas of central photopenia and increased peripheral uptake. Angiography illustrates that most GCTs are hypervascular lesions. CT scans demonstrate soft tissue attenuation with well-defined margins and a thin rim of sclerotic bone. MRI exhibits characteristic heterogeneous signal intensity with low-to-intermediate intensity on both T1- and T2-weighted images.

Most GCTs are benign; malignant GCTs occur in only 5% of cases. Malignant GCTs usually are related to previous irradiation in the vicinity of the tumour. Although most GCTs are benign, the lesions are locally aggressive, and their size and location may not allow complete resection. Those that cannot be excised en bloc should be curetted. Radiation is reserved for surgically inaccessible tumours. Selective arterial embolization also can be used in the management of these tumours. Recurrence rates can be as high as 40-60%.

**Primary Malignant Spinal Tumours**

Levine and Crandall offer a summary of the treatment of primary malignant tumours of the spine. For treatment of astrocytomas, a less common spinal tumour, see Minnehan et al. For treatment for non-ambulatory patients, see Kondo et al. Palliative surgery for metastatic thoracic and lumbar tumours is presented by Cho and Sung.

Kaloostian et al conducted a review of the literature regarding treatment and outcomes of patients with metastatic disease or primary tumours of the spinal column. They reported that en-bloc resection is the mainstay of treatment for malignant primary tumours of the spinal column, whereas intralesional resection is generally appropriate for benign primary tumours. Low-quality evidence supports the use of chemotherapy in select primary tumours. Radiation therapy is often used for incompletely resected or unresectable lesions.

According to this review, surgical considerations for the treatment of metastatic disease (see Metastatic Tumours in the Spine) are more nuanced and require consideration of patient performance status and the pathology of the primary tumour. Treatment of metastatic and primary tumours of the spinal column requires a multidisciplinary approach.

Glennie et al carried out a systematic review with consensus expert opinion regarding optimal reconstructive approaches after en-bloc resection of primary osseous spinal tumours. They reached the following conclusions:

- Posterior reconstruction with at least two vertebral levels above and below is recommended.
- Cages should be employed for single-level defects.
- Structural bone graft, either alone or in combination with a cage, should be employed for spanning a defect greater than two vertebral bodies.

The authors also noted that postoperative radiation therapy, if planned, may affect fusion strategy.

**Chondrosarcoma**

Chondrosarcoma is the second most common nonlymphoproliferative tumour of the spine. Chondrosarcomas make up 7-12% of all spine tumours, and the spine is the primary site in 3-12% of all chondrosarcomas. Men are affected two to four times more frequently than women. The mean age of presentation is 45 years. The thoracic spine is the most common...
site, but chondrosarcomas can occur at all levels of the spine. The most common symptoms are pain, a palpable mass, and neurologic complaints in 45% of patients.

Plain radiographs of chondrosarcomas typically demonstrate bone destruction. The lesions may be apparent in the vertebral body 15% of the time, in the posterior elements 40% of the time, or in both 45% of the time. In 70% of patients, the characteristic chondroid matrix in the form of rings and arcs is apparent on x-ray. Cortical destruction with soft-tissue extension is best observed on computed tomography (CT) or magnetic resonance imaging (MRI). (See the images below.)

Chondrosarcomas that arise from malignant transformation of osteochondromas are observed as a thickening of the cartilaginous cap. Involvement of the adjacent vertebral levels by extension through the disc is observed in 35% of all lesions. On CT or MRI, mineralization is usually apparent in the soft tissue component of the lesion. The radionuclide uptake by the lesion is intense and has a heterogeneous appearance on bone scan.
Chondrosarcomas are relatively low-grade lesions (grade I or II). Most lesions are primary chondrosarcomas rather than secondary chondrosarcomas that arise from the malignant degeneration of osteochondromas, as previously noted. Chondrosarcomas have relatively sparse cartilaginous stroma with a surrounding pseudocapsule. Examination under higher magnification reveals atypical nuclei with several mitotic figures per high-powered field.

Surgical resection by vertebral corpectomy and strut bone grafting sometimes may be necessary for complete excision. Cure is possible when complete resection can be achieved; this is possible 25% of the time. If wide marginal resection cannot be achieved, the tumour recurrence results in death in 74% of cases. The mean survival for all patients with chondrosarcomas is 5.9 years according to Shives et al. See the image below.

Adjunctive treatment with radiation is controversial for these tumours. Chemotherapy is used sometimes to help decrease the size of the mass with high-grade chondrosarcomas and dedifferentiated chondrosarcomas. Metastases of chondrosarcoma depend on the grade of the primary chondrosarcoma. The lungs are the most frequent sites of metastasis.

**Ewing sarcoma**

Ewing sarcoma is the most common nonlymphoproliferative primary malignant tumour of the spine in children. Lesions of the spine make up 3-10% of all primary sites of Ewing sarcoma. Metastatic foci of Ewing sarcoma involving the spine are more common than primary lesions of the spine. Patients with Ewing sarcoma usually present when aged 10-20 years.

The most common site of occurrence in the spine is the sacrococcygeal region followed by the lumbar and thoracic segments. Ewing sarcoma rarely occurs in the cervical spine. Lesions are centred primarily in the vertebral body but they can extend into the posterior elements.

Plain x-rays reveal permeative bone lysis, osseous expansion, or sclerosis. Diffuse sclerosis is observed in 69% of spinal lesions and is associated with osteonecrosis. CT and MRI demonstrate osseous involvement as well as surrounding soft tissue involvement. However, MRI is nonspecific.

Tissue from a Ewing sarcoma is composed of sheets of small, round, blue cells divided by septa, scant cytoplasm, and abundant collagen. Areas of osteonecrosis are found in spinal lesions. These correspond to the sclerotic areas observed on plain x-rays, as discussed.

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*Figure 32 Spinal Tumours - T2 weighted MRI scan of previous tumour*
above. Genetically, patients with Ewing sarcoma are found to have an 11;12 chromosomal translocation.

Before the advent of chemotherapy, the survival rate for patients with Ewing sarcoma was dismal because of the inability to completely resect these lesions, especially in the axial skeleton.

Radiation and chemotherapy are the current mainstays of treatment of Ewing sarcoma in the spine, achieving almost 100% local control with an 86% long-term survival rate in patients with spinal Ewing non-sacral sarcomas. Sacral tumours have a 62% local control rate and only 25% long-term survival rate because of the tendency for delayed clinical presentation and larger tumour size. The most important prognostic indicator for survival of Ewing sarcoma is the tumour's response to chemotherapy.

**Osteosarcoma**

Osteosarcomas of the spine are rare, making up only 0.6-3.2% of all osteosarcomas and only 5% of all primary malignant tumours of the spine. They typically present in patients in the fourth decade of life and have a male predominance. Osteosarcomas are found at all levels of the spine but are most common in the lumbosacral segments. Eccentric involvement of the vertebral body with extension into the posterior elements is common.

Patients often present with pain and a palpable mass. Neurologic symptoms, ranging from sensory deficits to paresis, are found in 70-80% of patients. Serum alkaline phosphatase may be elevated.

Plain x-rays of spinal osteosarcomas reveal a densely mineralized matrix, giving rise to the term ivory vertebrae. A loss of vertebral height often occurs, with sparing of the adjacent disc. Purely lytic lesions also have been described. CT and MRI are useful for evaluating the extent of bony and soft-tissue involvement. If a large amount of mineralized matrix is present, the lesion may appear with low signal intensity on all MRI sequences.

Most osteosarcomas are blastic lesions (osteoblastic, chondroblastic, or fibroblastic). Osteosarcomas can arise primarily or secondarily from an exposure to radiation. Secondary osteosarcomas can have a latency of up to 20 years. Spinal osteosarcomas also have been found in patients with Paget disease.

Surgical resection is the rule; however, resection of spine lesions is often incomplete due to the size and location of the tumour at the time of presentation. Adjuvant chemotherapy and radiation therapy often are employed with varying degrees of utility. Spinal osteosarcomas have a dismal prognosis, with deaths usually occurring within the first year of diagnosis. Only a few patients have been reported to survive longer than 2 years.

**Chordoma**

Luschka first described chordoma morphologically in 1856 in Virchow's lab. The discovery of the notochordal nature of the tumour and the coining of the term chordoma is credited to Ribbert in 1894.

Chordomas are uncommon, accounting for 2-4% of all primary malignant bone tumours with a prevalence of 0.51 per million. However, they are excluding lymphoproliferative tumours and metastases, the most common primary malignant tumour of the spine in the adult.

As Ribbert described, chordomas arise for the notochord remnant. The notochord normally evolves into the nucleus pulposus of the intervertebral discs. Non-neoplastic notochord vestiges also are found at the midline of the sphenoccipital synchondrosis and in the
sacrococcygeal regions. The locations in which chordomas occur parallel these vestigial distributions.

Regarding chordoma prevalence, 30-35% occur in the sphenoorcipital region, 50% in the sacrococcygeal region (especially S4-S5), and 15% occur in the other spinal segments.

Interestingly, chordomas have not been reported to arise from the intervertebral discs. Chordomas occur most commonly in patients aged 30-70 years, with a peak incidence in the fifth to sixth decades of life. Sphenoorcipital lesions have equal sex distributions but sacrococcygeal lesions have a 3:1 male-to-female ratio.

Presentation of chordomas is often subtle, with a gradual onset of pain, numbness, motor weakness, and constipation or incontinence. Constipation is a uniform finding in most patients with sacrococcygeal lesions. Chordomas are typically slow-growing lesions and are often very large at the time of presentation.

On plain x-ray, chordomas appear as a destructive lesion of a vertebral body in the midline, with a large associated soft tissue mass. In sacrococcygeal lesions, osseous expansion is frequent and may extend across the sacroiliac joints. Mineralization within the tumour may be observed on the plain x-rays of 50-70% of sacrococcygeal lesions. The mineralization is amorphous and predominates in the periphery of the lesion.

Lesions in spinal segments above the sacrum are less expansile and demonstrate evidence of calcification in only 30% of cases. They may have areas of sclerosis in 43-62% of cases. The intervertebral discs above or below a chordoma may be involved and narrowed in a manner that simulates infection. The lesion can make its way through the intervertebral disc to infiltrate an adjacent level. This occurs in approximately 11-14% of cases.

CT demonstrates both osseous and soft-tissue components of the tumour. Coronal and sagittal reconstructions of the CT scan are helpful in assessing neural foraminal and sacroiliac joint involvement. MRI is an important adjunct in the workup of chordomas. The lesions appear with low- to intermediate signal intensity on T1 images with very high signal intensity on T2 images, reflecting the high-water content of chordomas. Enhancement occurs following intravenous contrast on both CT and MRI.

Chordomas are lobulated neoplasms, which usually are contained within a pseudocapsule. Histology of these lesions reveals long cords of physaliphorous cells. Physaliphorous cells are clear cells containing intracytoplasmic vacuoles with abundant intracellular and extracellular mucin. Sarcomatous chondroid, osteoid, or fibroid elements may be found within the chordoma.

Surgical resection is the rule. Adjuvant postoperative radiation therapy, proton beam therapy, and brachytherapy all have been used with varying results. The prognosis depends on whether the tumour can be resected completely. The location of the lesion and the size at presentation often necessitate incomplete resection. The treatment of sacral chordoma is an arduous clinical undertaking that requires a multidisciplinary approach and attention to detail from the outset.

Despite aggressive, well-planned surgical management and adherence to strict surveillance protocols, frequent recurrence and the late onset of metastatic disease are to be expected in a substantial proportion of patients, especially those with a large chordoma or one at a more cephalad level. Adequate surgical treatment results in substantial functional impairment and numerous complications; however, it does offer the possibility of long-term disease-free survival.
Persons with sacrococcygeal tumours often have improved survival because the surrounding structures are relatively more expendable and allow a more complete resection. Persons with sacrococcygeal lesions typically have 8-10 years survival, as opposed to 4-5 years survival for persons with chordomas in other spinal sites. Death usually is related to local recurrence and invasion rather than metastatic disease. Chordomas can metastasize. The most common sites of metastases are the liver, lungs, regional lymph nodes, peritoneum, skin, and heart.

**Multiple myeloma**

Multiple myeloma is a systemic disease that affects middle-aged people and is characterized by areas of local bone destruction. Multiple myeloma is the most common primary malignancy of bone and the spine. The underlying cell line is the malignant plasma cell, which produces abnormal quantities of immunoglobulins.

The presentation of patients with myeloma is similar to that of other spine tumour patients. Patients complain of pain that may be worse at night. The laboratory workup for these patients should include a complete blood count with differential looking for anaemia and thrombocytopenia, an elevation of the erythrocyte sedimentation rate, and a decrease in the serum albumin with increased total serum protein. The abnormal production of immunoglobulins can be detected on serum or urine electrophoresis and can be used to confirm the diagnosis.

Radiographically, skeletal survey is used to screen for lesions that can occur throughout the skeleton. Bone scans have a high false-negative rate and are not optimal studies for the evaluation of myeloma. Once a lesion is detected in the spine, CT, MRI, or both should be performed to assess the destruction of the vertebrae and the effect of this destruction on the surrounding neurologic and paraspinal tissues.

Multiple myelomas are generally sensitive to radiation therapy and chemotherapy. Surgery for stabilization is indicated in myelomas of the spine when destruction of the vertebral body exists to such an extent that collapse and possible kyphosis with canal compromise could result.

Prophylactic posterior stabilization can be carried out with segmental instrumentation in cases prior to fracture. Anterior strut grafting or cage reconstruction may be necessary once fracture and collapse have occurred. Adjuvant radiation therapy may be used postoperatively once healing of the surgical site has been obtained.

**Solitary plasmacytoma**

Akin to multiple myeloma as a descendent of plasma cell malignancies, plasmacytoma is a solitary lesion that usually affects the vertebral body. Plasmacytomas generally affect younger patients than multiple myeloma and are associated with a better prognosis. Plasmacytomas eventually can evolve into multiple myeloma; thus, patients should be monitored for more than 20 years following the original diagnosis of plasmacytoma.

The diagnosis is made by biopsy of the lesion, and treatment includes radiation and bracing except in persons with pathologic or impending pathologic fractures. In these individuals, surgical resection and stabilization should be carried out with postoperative adjuvant radiation therapy once 6-8 weeks of postoperative healing has occurred. Patients have greater than 60% 5-year survival rates.

**Metastatic Tumours in the Spine**

The tumours that most commonly metastasize to the spine are as follows:
Tatsui et al found that patients with prostate cancer had the highest rate of metastases to the spine. They also found that lung cancer was the most common primary lesion in patients whose spinal metastases were detected before the diagnosis of primary lesions.

The time from diagnosis of the primary lesion to detection of the spinal metastasis was shown to be shortest in patients with lung cancer and longest in those with breast cancer. Patients with metastases from breast cancer or prostate cancer had the highest 1-year survival rate, whereas patients with metastases from lung or gastric cancer had the lowest 1-year survival rates.

Advances in the molecular understanding of metastatic spinal tumours are likely to bring about substantial changes in management paradigms. Through molecular characterization, critical information can be obtained that can be used for making the initial diagnosis, determining the optimal treatment approach, evaluating the efficacy of treatment, and monitoring for recurrence, as well as for predicting complications, clinical outcome, and overall survival.

Complications

Complications associated with spinal tumours can be divided into the following two groups:

- Complications associated with the tumour, its recurrences, or its metastases - Neurologic complications include radicular pain or focal weakness from impingement on a nerve root and complete or incomplete paraplegia from direct pressure on the spinal cord.
- Complications associated with surgical, radiation, or chemotherapeutic treatment of the tumours - Complications that result from the treatment modality employed may be related to structures sacrificed during the surgical resection to obtain clear margins, structures in the path of radiation therapy, or the systemic effects of chemotherapy.

Fan et al conducted a study to analyze complications following posterior vertebral column resection in patients with spinal tumours. A total of 36 complications were reported, and the following associations were noted:

- Transient late tracheal extubation was associated with higher intraoperative bleeding volume and lower preoperative forced vital capacity and forced expiratory volume in 1 sec.
- Subsidence in the replaced spinal segment was associated with increased duration of surgery, higher intraoperative bleeding volume, and higher total blood transfusion volume.
- Thrombocytopenia was associated with increased duration of surgery and higher total blood transfusion volume.

The majority of the complications were minor and did not affect patient recovery. The investigators concluded that active preventive measures are necessary to reduce the incidence of major complications.
Outcomes
Chang et al conducted a study to evaluate local control rate and to identify prognostic factors after stereotactic radiosurgery for treatment of primary malignant spinal tumours. Median age of the 29 patients was 46 years (range, 11-68). Histologic diagnoses included chordoma (n=1), chondrosarcoma (n=5), osteosarcoma (n=3), synovial sarcoma (n=3), plasmacytoma (n=2), Ewing sarcoma (n=2), malignant peripheral nerve sheath tumour (n=2), and malignant fibrous histiocytoma (n=1). Mean follow-up was 50 months (range, 8-126).

Surgical resection was the initial treatment in 25 cases, percutaneous biopsy in four. Stereotactic radiosurgery was used as primary treatment in 14 cases and as salvage treatment for progressive lesions in 15. Eleven patients had undergone previous conventional external-beam radiation therapy before stereotactic radiosurgery. Median tumour volume was 14 cm$^3$ (range, 2.0-235). Delivered radiation doses were 12-50 Gy in two to six sessions. The mean radiation dose converted into a biologic effective dose (BED) was 60 Gy (range, 43-105).

Mean overall survival was 84 months for chordoma patients and 104 months for sarcoma patients. The investigators found no factors that affected overall survival. The mean local progression-free survival was 56 months for chordoma patients and 73 months for sarcoma patients. The recurrent mode of presentation was predictive of local progression of spinal sarcomas. For patients with chordoma, no factors were found to correlate with local recurrence.

Kose et al conducted a study of the effect of early rehabilitation on neurofunctional outcome after surgery in children with spinal tumours. The investigators reviewed medical charts and radiographic records of 70 paediatric patients (aged 1-17 years) who underwent surgery for the removal of spinal tumour. The patients received rehabilitation treatment beginning 4 days (range, 2-7 days) after surgery for 10 days (range, 7-23 days).

Results were assessed on the basis of scoring on the Modified McCormick Scale, the Functional Independence Measure for Children, the American Spinal Injury Association Impairment Scale, and the Karnofsky Performance Status Scale. Sensory function, motor function, and activity of daily living were significantly improved for the patients who received early rehabilitation. Tumour setting, the level of localization, and the patients' clinical symptoms had no bearing on neurofunctional outcomes.

Paget Disease
Paget disease of bone (osteitis deformans) is a metabolic disorder characterized by abnormal osseous remodelling. Sir James Paget first described Paget disease in 1877 as a chronic inflammatory remodelling disease of bones. He termed the condition osteitis deformans. Paget disease of bone is caused by a localized increase in osteoclastic and osteoblastic activity and can progress to involve the entire bone. Deformed, enlarged bones is a common feature, especially in weight-bearing areas. Genetic factors play an important role in pathogenesis, with evidence that susceptibility may be determined by variants in or near genes that regulate osteoclast function.

Paget disease is depicted in the images below.
Fig 47 is a radiograph of the tibia in a patient with Paget sarcoma reveals a destructive bone-forming mass in the proximal tibia (osteosarcoma).

Fig 48 Shows a skull radiograph in a patient with Paget disease demonstrates a large, well-circumscribed lytic lesion (arrows) in the frontal and parietal bones (osteoporosis circumscripta).

Fig 49 shows the first sacral vertebra and demonstrates marked cortical thickening (arrows) and trabecular coarsening.
Fig 50 shows the knee in a patient with Paget disease reveals prominent dark lines in the medullary bone, indicating trabecular coarsening (arrows).

Paget disease evolves through 3 stages:

1. An early lytic or hot phase;
2. An intermediate or mixed phase
3. A final or cold phase, marked by dense bone formation

Paget disease rarely is diagnosed in the initial lytic phase. At this early point of the disease, osteoclastic activity is predominant. Paget disease usually begins at the end of a bone, except when it occurs in the tibia. A characteristic sharply demarcated zone of osteolysis may begin in the subcortical bone and advance along the diaphysis. Osteoblastic activity lags behind; thus, radiolucent fibrous tissue replaces normal bone.

The intermediate or mixed phase reveals evidence of osteolytic and disorganized osteoblastic activity. New bone forms abnormally and demonstrates characteristically coarsened trabecula and cortical thickening in the cancellous and compact bone, respectively. Characteristic intracytoplasmic inclusions may be observed microscopically, supporting evidence for the viral aetiology theory.

The final or cold phase demonstrates less evidence of continual osseous remodelling. Previously laid down woven bone is converted to dense lamellar bone. Histologic features of disorganized bone are prominent. The intersecting lines of remodelled bone have a characteristic mosaic pattern histologically.

Insufficiency fractures in patients with Paget disease may present with pain that can last up to several weeks. If pain is focal and severe, it may be a sign of an impending, complete fracture, and radiographic evaluation is warranted. Insufficiency fractures most frequently affect the femur and tibia. Involvement in critical weight-bearing locations may lead to fracture or severe secondary arthritis. See the images below.

Fig 51 shows the hip in a patient with Paget disease demonstrates dense sclerosis involving the femoral head and neck (arrows). This is a high-risk area for insufficiency fracture.
Preferred examination

The radiographic findings of Paget disease are diagnostic in many patients. The lytic stage most commonly is observed in the skull and long bones. The typical appearance in the long bones is osteolysis, which begins in the epiphysis and advances along the diaphysis. Trabecular coarsening and distortion and cortical thickening are observed in the sclerotic phase, typically involving the axial skeleton.

Radiographic findings in Paget disease often are pathognomonic, particularly in the lytic phase. However, given the variable imaging appearance of Paget disease in different stages, as well as the many different bones involved, the differential diagnosis may vary substantially among patients.

Radiography

In the skull, the lytic phase (osteoporosis circumscripta) typically involves the frontal or occipital bones and progresses to a mixed pattern with multifocal sclerotic patches in the intermediate stage of the disease, referred to as a cotton wool appearance. See the images below.

*Figure 38 A-P X-Ray of femur*

Fig 52 shows the femur in a patient with late-stage Paget disease reveals a transverse insufficiency fracture through the proximal femoral shaft (banana fracture).

*Figure 39 Paget Disease - Lateral X-Ray of calvarium*

Fig 53 Shows the calvarium in a patient with Paget disease reveals multiple patches of sclerotic bone in the calvarium (cotton wool appearance).
The vertebral bodies typically become enlarged with a prominent cortical margin (picture frame vertebrae) or become densely sclerotic, mimicking lymphoma or metastatic disease (ivory vertebra). See the images below.

Fig 54 shows a lumbar spine in a patient with Paget disease demonstrates enlargement of an involved vertebral body (arrow), with sclerosis more prominent at the vertebral endplates (picture frame vertebra).

Fig 55 shows the upper thoracic spine and reveals a densely sclerotic vertebral body (ivory vertebra) caused by Paget disease (arrow). The appearance mimics findings that can be observed with malignant neoplasm such as lymphoma or metastatic disease.

In the pelvis, typical findings include thickening of the iliopsoas line in early stages, progressing to patchy sclerosis and lucency in later stages. See the image below.

Fig 56 shows a pelvis and demonstrates thickening of the right iliopsoas line (small arrows), as well as later-stage involvement with Paget disease, including trabecular coarsening and patchy sclerosis (large arrow).

Weakening of the pagetic acetabular bone may lead to protrusio acetabuli and insufficiency fracture. See the image below.
Fig 57 shows a radiograph of the right hip, coned down on the obturator ring, reveals patchy sclerosis and disorganized coarsened trabecula characteristic of late-stage Paget disease. An insufficiency fracture of the ischium inferior to the acetabulum (arrows) is present.

In the long bones, early involvement consists of lysis of the subarticular bone, which advances along the diaphysis with the characteristic shape of a blade of grass. Long bones are affected first in the epiphyseal region, with the exception of the tibia, where Paget disease frequently begins in the tubercle.

Later stages of disease show development of enlarged, sclerotic, deformed bones with thickened coarse trabeculae. The weakened femur and tibia eventually may become bowed under the stress of weight bearing. Insufficiency fractures may occur, characteristically involving the convex cortical surface. Conversely, Looser zones of osteomalacia typically occur on the concave cortical surface. See the images below.

Fig 58 shows a coned down anteroposterior radiograph of the knee demonstrates an abnormal lucency in the distal femur with a flame-shaped or "blade of grass"-shaped proximal margin caused by the advancing lytic phase of Paget disease (black arrows).
Fir 59 shows a coned down lateral radiograph of the tibia reveals replacement of the tibial tubercle with pagetic bone that has mixed osteolysis and sclerosis (arrows).

Fig 60 shows an anteroposterior radiograph of the distal forearm demonstrating mixed-phase Paget disease in the distal radius with lytic disease proximally (white arrows) and coarsened trabeculae more distally (black arrow).

Fig 61 shows an anteroposterior radiograph of the proximal femur involved with intermediate (mixed phase) Paget disease and reveals characteristic insufficiency fractures on the convex surface of the bone (arrow).

Development of secondary sarcoma in pagetic bone is the most lethal complication of Paget disease, occurring in 1% or fewer of patients with Paget disease (see the image below). These sarcomas are aggressive and may be multicentric. Short-term interval follow-up and/or cross-sectional imaging may prevent diagnostic errors and initiate prompt attention to a newly developing lesion.
Fig 62 shows a radiograph of the tibia in a patient with Paget sarcoma reveals a destructive bone-forming mass in the proximal tibia (osteosarcoma).

**Computed Tomography**

Cross-sectional MRI and CT demonstrate the altered bone structure seen in Paget disease, such as coarsened trabeculae, thickened cortices, and bone hypertrophy. Findings that are present on plain radiography are often better displayed on CT. MRI does not depict the changes in mineralization as well as traditional radiography or CT. MRI, however, can show alterations in marrow characteristics that can mirror the pathologic changes occurring during the course of the disease. Thus, earlier in the disease, marrow signal can be normal or hypervascular, while later changes can show fatty replacement of the marrow.

The anatomy is well demonstrated by cross-sectional imaging in complex structures, such as the spine, where spinal or nerve root compression may be an issue. Cross-sectional imaging also helps delineate the pathology in complicated Paget disease, which includes nerve or spinal cord compression, as well as basilar invagination at the skull base and osseous encroachment involving cranial nerve foramina.

Secondary sarcomatous development also is better evaluated with cross-sectional imaging. Additionally, should biopsy be indicated for the diagnosis of sarcoma, CT typically is the guidance modality of choice. See the images below.

Fig 63 shows the first sacral vertebra and demonstrates marked cortical thickening (arrows) and trabecular coarsening.
Fig 64 shows a sagittal CT image of the cervical spine and demonstrates increased sclerosis within the C5 vertebral body. This is the CT version of the "ivory vertebra". Also note the subtle thickening of the cortex as compared with the adjacent vertebral bodies.

Fig 65 shows an axial T1-weighted MRI of the knee in a patient with Paget disease and reveals prominent dark lines in the medullary bone, indicating trabecular coarsening (arrows).

Fig 66 shows a sagittal T1-weighted MRI of the lumbar spine and demonstrates the enlargement of the fourth lumbar vertebral body with no central canal encroachment (arrows).
Fig 67 shows a sagittal T1-weighted MRI of the lumbosacral spine and demonstrates Paget disease in the sacrum. There is increased signal intensity throughout the sacral marrow, indicative of fatty replacement. Compare with the marrow signal characteristics of the lumbar vertebrae, as well as the adjacent intra-pelvic fat.

Fig 68 shows a coronal T1-weighted MRI of the hips and demonstrates Paget disease of the proximal left femur. Note the coarsened trabeculae and thickened cortex, changes which are low signal on MRI.

**Nuclear Imaging**

Skeletal scintigraphy is useful. Radionuclide bone scans are more sensitive than radiographs for the diagnosis of Paget disease. Additionally, bone scans help survey the different sites of involvement with polyostotic disease.

Characteristically, a marked uptake of radiopharmaceutical in the involved bones is observed. However, late-stage involvement may not reveal intense radiopharmaceutical uptake, and osteoporosis circumscripta may demonstrate only a peripheral rim of increased uptake. Scintigraphy tends to follow the physiologic activity of disease and may monitor treatment.
Polyostotic Paget disease often can be distinguished from multiple metastatic lesions, although occasional difficulties occur. Perform radiographic correlation when this situation arises. Furthermore, the diagnosis of fracture or sarcoma may be challenging, often requiring multimodality correlation. See the images below.

Fig 69 shows a whole-body bone scan in a patient with polyostotic Paget disease reveals intense uptake of radiopharmaceutical in the femur, pelvis, spine, and proximal right humerus. The cortical discontinuity of the proximal right humerus represents an insufficiency fracture (arrow).

Fig 70 shows the anterior image of the thoracic and lumbar spine in a 75-year-old man and demonstrates intense radiopharmaceutical uptake in the third lumbar vertebra, which is involved with Paget disease (arrows)

Approach Considerations
The short-term objective of Paget disease treatment is to control disease activity. The long-term objectives of treatment are to minimize or prevent disease progression and to decrease complications from the disease, if possible.

Indications for drug treatment of Paget disease are as follows:

- Metabolic active disease - Bone pain, fatigue fracture, skull/spine fracture, radiculopathy, osteolytic lesions, bony deformities, weight-bearing bone involvement, compression of spinal cord and nerve roots, bone compression of the eighth cranial or optic nerve
• Preparation for orthopaedic surgery (joint replacement anticipated at involved sites within 6 months)
• Hypercalcemia or hypercalciuria - Recurrent renal calculi due to hypercalciuria, or fractures; serum alkaline phosphatase or urine hydroxyproline levels greater than twice the upper limit of the reference range; immobilization

When Paget disease occurs around a joint, treatment is often administered in an attempt to prevent development of osteoarthritis. In addition, young patients with Paget disease and those with high levels of bone-specific alkaline phosphatase (BSAP) are often treated to avoid future complications. The concept that aggressive treatment is associated with prevention of progression and reduction in risk of future complications is not yet supported by clear findings from long-term placebo-controlled trials; however, indirect evidence suggests that this hypothesis is reasonable.

Drug therapy for Paget disease should include bisphosphonate treatment with serial monitoring of bone markers. Response to therapy is indicated by reduction of symptoms and decreases in levels of BSAP (a bone formation marker) and deoxypyridinoline, C-telopeptide, or N-telopeptide (bone resorption markers).

Secondary osteoarthritic pain may be reduced by nonsteroidal anti-inflammatory drugs or other nonnarcotic analgesics. In contrast, bone pain in Paget disease typically responds poorly to these pain medications. Patients should receive 1000-1500 mg of calcium and at least 400 U of vitamin D daily. This recommendation is especially important in conjunction with bisphosphonate treatments.

Orthotic devices, including canes and walkers, may be useful for patients with gait abnormalities resulting from Paget disease that involves the lower limbs.

Because of the increased risk of malignancy, patients with Paget disease should be monitored indefinitely. Chemotherapy, radiation, or both may be used to treat neoplasms that arise from pagetic bone. Amputation may also be necessary in the presence of a malignant transformation.

**Torticollis**

*Torticollis*, also known as wry neck or loxia, is a dystonic condition defined by an abnormal, asymmetrical head or neck position, which may be due to a variety of causes. The term *torticollis* is derived from the Latin words *tortus* for twisted and *collum* for neck.

It can appear in various forms:

- Congenital
- Spasmodic
- Acquired

Figure 57 Torticollis
Congenital

The aetiology of congenital muscular torticollis is unclear. Birth trauma or intrauterine malposition is considered to be the cause of damage to the sternocleidomastoid muscle in the neck. This results in a shortening or excessive contraction of the sternocleidomastoid muscle, which curtails its range of motion in both rotation and lateral bending. The head typically is tilted in lateral bending toward the affected muscle and rotated toward the opposite side. It can be an expression of upset at the cranial base, not only at the occipitoatlasantal joint, but tension patterns affecting the junction of the occiput and temporal bone, with the jugular foramen between the two (from whence emerges the ascending spinal accessory nerve) supplying sternocleidomastoid and trapezius.

The reported incidence of congenital torticollis is 0.3-2.0%. Sometimes a mass, such as a sternocleidomastoid tumour, is noted in the affected muscle at the age of two to four weeks. Gradually it disappears, usually by the age of eight months, but the muscle is left fibrotic.

Initially, the condition is treated with physical therapies, such as stretching to release tightness, strengthening exercises to improve muscular balance, and handling to stimulate symmetry. A TOT Collar is sometimes applied. About 5–10% of cases fail to respond to stretching and require surgical release of the muscle.

Spasmodic torticollis

This is a torticollis with recurrent, but transient contraction of the muscles of the neck and especially of the sternocleidomastoid. Synonyms are "intermittent torticollis", "cervical dystonia" or "idiopathic cervical dystonia", depending on cause.

Acquired torticollis

Non-congenital muscular torticollis may result from scarring or disease of cervical vertebrae, adenitis, tonsillitis, rheumatism, enlarged cervical glands, retropharyngeal abscess, or cerebellar tumours. It may be spasmodic (clonic) or permanent (tonic). The latter type may be due to Pott's disease (tuberculosis of the spine).

- A self-limiting spontaneously occurring form of torticollis with one or more painful neck muscles is by far the most common ('stiff neck') and will pass spontaneously in 1–4 weeks. Usually the sternocleidomastoid muscle or the trapezius muscle is involved. Sometimes draughts, colds, or unusual postures are implicated; however in many cases no clear cause is found. These episodes are commonly seen by physicians.
- Tumours of the skull base (posterior fossa tumours) can compress the nerve supply to the neck and cause torticollis, and these problems must be treated surgically.
- Infections in the posterior pharynx can irritate the nerves supplying the neck muscles and cause torticollis, and these infections may be treated with antibiotics if they are not too severe, but could require surgical debridement in intractable cases.
- Ear infections and surgical removal of the adenoids can cause an entity known as Grisel's syndrome, a subluxation of the upper cervical joints, mostly the atlantoaxial joint, due to inflammatory laxity of the ligaments caused by an infection. This bridge must either be broken through manipulation of the neck, or, surgically resected.
- The use of certain drugs, such as antipsychotics, can cause torticollis.[9]
- Antiemetics - Neuroleptic Class - Phenothiazines
There are many other rare causes of torticollis.

**Spondylopathies**

In medicine, *spondylopathies* is a general term for disorders of the vertebrae. The word derives from *spondylos* (vertebra) and *pathos* (suffering). When involving inflammation, it can be called *spondylitis*. In contrast, a *spondyloarthropathy* is a condition involving the vertebral joints, but many conditions involve both spondylopathy and spondyloarthropathy.

Conditions like this include:

- Ankylosing Spondylitis
- Spondylosis

**Ankylosing Spondylitis**

Ankylosing *spondylitis* (Greek *ankylos*, fused; *spondylos*, vertebra; *-it is*, inflammation), previously known as *Bechterew's disease* (or syndrome) and *Marie-Strümpell disease*, is a chronic inflammatory disease of the axial skeleton, with variable involvement of peripheral joints and non-articular structures.

It is a member of the group of the spondyloarthropathies, with a strong genetic predisposition. It mainly affects joints in the spine and the sacroiliac joint in the pelvis. In severe cases, it can eventually cause complete fusion and rigidity of the spine.

"Bamboo spine" develops when the outer fibres of the fibrous ring of the intervertebral disks ossify, which results in the formation of marginal syndesmophytes between adjoining vertebrae.

*Figure 58 Ankylosing Spondylitis beside normal spine*
The pictures above show a normal spine beside a spine with ankylosing spondylitis. Note:

- The absence of any outline of the sacroiliac joints
- The absence of any disc space, as outer fibres of the intervertebral disc has ossified, causing the characteristic ‘Bamboo spine’ effect, see here

**Symptoms**

- Symptoms appear gradually, most commonly between 15 and 45 years of age, more often in males
- Since the initial signs and symptoms are not specific for ankylosing spondylitis, there is a lag-time between onset of disease and diagnosis, which averages between 8.5 years and 11.4 years
- Sacroiliitis is usually one of its earliest manifestation.
- The initial symptom is usually a typical chronic dull pain, insidious in onset, felt deep in the lower lumbar or gluteal region, accompanied by low-back morning stiffness. It can occur in in the middle part of the spine or the entire spine, often with pain referred to one or the other buttock or the back of the thigh from the sacroiliac joint. This pain is often severe at rest, but improves with physical activity. However, many experience inflammation and pain to varying degrees regardless of rest and movement.
- As the inflammation spreads up the spine, the costovertebral joints can also be involved, with pain on deep breathing and a reluctance to move into extension. This can result in a significant kyphosis before the spine fuses. The end result can be a straight lumbar spine and a kyphotic upper thoracic spine
- Loss of spinal mobility, with limitation of anterior flexion, lateral flexion, and extension of the lumbar spine, is seen with chest pain and generalized fatigue
- The most serious complication of the spinal disease is spinal fracture, which can occur with even minor trauma to the rigid, osteoporotic spine.
- Arthritis in the hips and shoulders may occur.
- The most common extra-articular manifestation is acute anterior uveitis which can antedate the spondylitis. About 40 percent of AS patients experience inflammation in the anterior chamber of the eye (uveitis), causing redness, eye pain, floaters and photophobia. Visual acuity is usually maintained and the fundus is normal. This is thought to be due to the association that both AS and uveitis have with the inheritance of the HLA-B27 antigen.
Pulmonary involvement, is characterized by slowly progressive apical lung fibrosis. There is limitation of chest expansion. Recurrent chest infection is the most common cause of death.

Aortitis, aortic valve insufficiency or cardiac conduction disturbances.

Prostatitis occurs with increased frequency in men.

Mortality attributable is largely the result of spinal trauma, aortic insufficiency, respiratory failure, amyloid nephropathy, or complications of therapy such as upper gastrointestinal haemorrhage.

When the condition presents before the age of 18, it is relatively likely to cause pain and swelling of large limb joints, particularly the knee. In prepubescent cases, pain and swelling may also manifest in the ankles and feet, where calcaneal spurs may also develop.

It is a systemic rheumatic disease, meaning it affects the entire body. Approximately 90% of AS patients express the HLA-B27 genotype, meaning there is a strong genetic association. 1-2% of individuals with the HLA-B27 genotype contract the disease. Tumour necrosis factor-alpha (TNF α) and IL-1 are also implicated in ankylosing spondylitis (both are cytokines of the acute inflammatory reaction).

As there is no cure, treatment usually focusses on pain reduction medication and physiotherapy.

**Spondylosis**

*Spondylosis* (usually called ‘wear and tear’ of the disc, or disc degeneration, is an old term (no longer used) referring to degenerative osteoarthritis of the joints between the centre of the spinal vertebrae and/or neural. If this condition occurs in the zygapophyseal joints, it can be considered facet syndrome, or spondylarthritis. If severe, it may cause pressure on nerve roots with subsequent sensory and/or motor disturbances, such as pain, paraesthesia, or muscle weakness in the limbs.

It is seen as a degenerative condition, that the discs have a structure that keeps the vertebral bodies apart, but with time there is loss of the disc space. It is seen as wear and tear, or degenerative, as it usually occurs later in life. It usually manifests as chronic, sometimes disabling, low back or neck pain (depending upon the area affected). Pain such as this is usually treated with pain medication, but if persists further examination is called upon in the form of X-ray or MRI (see right). The x-ray usually reveal narrowing off the disc space and the presence of osteophytes (outgrowths of bone around the periphery of the vertebral body).

When the space between two adjacent vertebrae narrows, compression of a nerve root emerging from the spinal cord may result in radiculopathy (sensory and motor disturbances, such as severe pain in the neck, shoulder, arm, back, and/or leg, accompanied by muscle
weakness). Less commonly, direct pressure on the spinal cord (typically in the cervical spine) may result in myelopathy, characterized by global weakness, gait dysfunction, loss of balance, and loss of bowel and/or bladder control. The patient may experience a phenomenon of shocks (paraesthesia) in hands and legs because of nerve compression and lack of blood flow. If vertebrae of the neck are involved it is labelled cervical spondylosis. Lower back spondylosis is labelled lumbar spondylosis.

**Causes of Spondylosis**
It is seen as a degenerative condition, but it is not really known what causes it.

Normally there is compression of the disc with weight bearing and this can lead to some loss of disc space over the course of a day. This compression can be relieved by lying down (in bed at night). Hence we are probably taller in the morning than we are at night.

It might be good to remind ourselves of the oblique and deepest muscles of the spine here. One way of looking at a possible cause of such ‘wear and tear’ is to see a chronic tension in these groups of muscles (and more particularly the fascia around these muscles). It can result in a chronic compression along the axis of the spine with a consequent compression of the intervertebral discs. Such a tension will not be relieved by rest or lying down. It manifests as chronic pain that may, or may not be relieved by pain-killers. The X-rays taken demonstrate bony changes and the characteristic osteophyte formation. Osteophytes are bony outgrowths that form at the periphery of the vertebral bodies and can be seen in the pictures above. They are diagnostic of the condition, but to see these suggests that such a condition has been present for 5-10 years.

Another way of seeing it is again via fascial tension patterns. Here though they can express themselves over regions of the spine allowing one or two levels to move. This creates what could be seen as a ‘hinge point’, with the adjacent bones touching off each other more readily. Such ‘touching’ will again create bony changes with osteophyte formation.

**Spondylolisthesis**
Spondylolisthesis is defined as forward translation of a vertebral body with respect to the vertebra below. The term is derived from the Greek roots spondylos, meaning spine, and listhesis, meaning to slide down a slippery path.

Spondylolisthesis can occur at any level of the spinal column, although it is most common in the lower lumbar spine. Most cases are thought to result from minor overuse trauma, particularly repetitive hyperextension of the lumbar spine. Spondylolysis, a break in the vertebra typically in the region of the pars interarticularis, may or may not be associated with a spondylolisthesis. If the pars defect is bilateral, it may allow slippage of the vertebra, typically L5 on S1, resulting in spondylolisthesis.
Both spondylolysis and spondylolisthesis are often asymptomatic, and the degree of spondylolisthesis does not necessarily correlate with the incidence or severity of symptoms, even when a patient is experiencing back pain. However, these 2 entities have been reported to be the most common underlying causes of persistent low back pain among children and adolescents, despite the fact that most cases are asymptomatic.

Spondylolisthesis can be classified into the following 6 distinct categories.

- **Type I**
  - Congenital (dysplastic)
  - Caused by agenesis of the superior articular facet

- **Type II**
  - Isthmic (spondylolytic)
  - Caused by pars interarticularis defects

- **Type III**
  - Degenerative
  - Secondary to articular degeneration

- **Type IV**
  - Traumatic
    - Caused by fracture or dislocation of the lumbar spine, not involving the pars

- **Type V**
  - Pathologic
    - Caused by malignancy, infection, or other types of abnormal bone

- **Type VI**
  - Postsurgical (iatrogenic)

*Figure 62 Spondylolisthesis at lumbosacral junction*
Epidemiology

Frequency

United States
The prevalence rate of isthmic spondylolisthesis is approximately 5% at age 5-7 years, with an increase to 6-7% by age 18 years. This condition is twice as common in males as in females, and the prevalence is lower in blacks (2.8%, black men; 1.1%, black women) than in whites (6.4%, white men; 2.3%, white women). Despite the higher prevalence in males, progression, although still rare, has been reported to be more common in females. Additional risk factors include having a first-degree relative with a slip, occult spina bifida at S1, and the presence of scoliosis.

Functional Anatomy

Mechanical stresses play an important role in this process. Erect posture produces a constant downward and forward thrust on the lumbar vertebrae. Stresses on the pars interarticularis are accentuated during repetitive hyperextension, which results in increased contact of the caudal edge of the L4 inferior articular facet with the L5 pars interarticularis. This collective trauma may eventually result in a stress fracture of the pars interarticularis. Spondylolisthesis may occur when bilateral pars defects are present, which allows forward slippage of the vertebra (typically L5 on S1). Spondylolisthesis has never been reported in quadrupeds or people who are chronically bedridden.

![Figure 63 Spondylolisthesis at L4/5](image)

Sport-Specific Biomechanics

Sports that involve repetitive hyperextension and axial loading of the lumbar spine may result in repetitive microtrauma to the pars interarticularis, resulting in spondylolysis and sometimes spondylolisthesis. Examples of such activities include gymnastics, football (lineman), wrestling, weightlifting (particularly standing overhead presses), rowing, pole vaulting, diving, hurdling, swimming (especially the butterfly stroke), baseball (especially pitching), tennis (especially serving), sailing (particularly the hiking manoeuvre), and volleyball. Gymnastics and football are generally considered the highest risk sports.
History

Typical findings when obtaining the history from a patient with spondylolisthesis may include the following:

- The patient is usually asymptomatic.
- The onset usually occurs during the growth spurt in late childhood and early adolescence, probably due to increased participation in strenuous sports during this period.
- Spondylolisthesis is an unlikely cause of back pain in adults (especially after age 40 y) with no history of symptoms before age 30 years; usually, another cause is identified (e.g., disc, strain).
- Low back pain is the usual symptom reported, and it is often exacerbated by motion, particularly lumbar extension and twisting. Radiation of pain into the buttocks is not uncommon. The patient may report relief of pain with extended periods of rest.
- Rarely, associated leg pain is present in the L5 or S1 distribution as a result of nerve root compression.
- Symptoms are often more severe during the advanced months of pregnancy.

Physical Examination

Findings noted during the physical examination may include the following:

- With high-grade slips, a palpable step-off may be felt over the spinous process at the level above the slipped vertebra because the posterior arch of the forward translated vertebra remains in place.
- Tenderness to deep palpation of the spinous process above the slip (typically L4) may be present. This palpation occasionally causes radicular pain.
- A positive one-leg hyperextension test (stork test) suggests a diagnosis of spondylolysis, but it is a nonspecific test with low sensitivity and low specificity.
- Hamstring tightness that is associated with all grades of symptomatic spondylolisthesis (see Grading) occurs at a rate of 80%. It commonly results in an abnormal gait, typically waddlileike, due to the inability of the patient to flex the hip with the knees extended.
- Paraspinal muscle spasm and tenderness are usually present.
- In advanced cases, a relatively short torso with a low rib cage, high iliac crests, and heart-shaped buttocks are noted.
- Limited forward flexion of the trunk is common with reduced straight-leg raising, which may cause pain but rarely any signs of nerve root tension.
- Postural deformity and a transverse abdominal crease are seen as a result of the pelvis being thrust forward.
- A thorough neurologic evaluation should be performed, including sensation in the sacral region to check for cauda equina compression.
- Weakness in the tibialis anterior muscle (L4 nerve root) is common.

Lumbosacral Spondylolisthesis Differential Diagnoses

Diagnostic Considerations

Diagnostic considerations include:

- Discogenic
- Infectious (discitis, osteomyelitis)
Mechanical low back pain (acute or chronic musculotendinous or ligamentous injuries, overgrowth syndrome, postural deformities)

Neoplastic (osteoid osteoma, aneurysmal bone cyst, chondroblastoma)

Spondylolysis / spondylolisthesis (acute [rare] vs chronic)

Vertebral growth plate injuries (growth plate fractures, Scheuermann’s)

Differential Diagnoses

- Degenerative Lumbar Disc Disease in the Mature Athlete
- Lumbar Disk Problems in the Athlete
- Lumbosacral Disc Injuries
- Lumbosacral Discogenic Pain Syndrome
- Lumbosacral Facet Syndrome
- Lumbosacral Radiculopathy
- Lumbosacral Spine Acute Bony Injuries
- Lumbosacral Spine Sprain/Strain Injuries
- Lumbosacral Spondylolysis
- Myofascial Pain in Athletes
- Pars Interarticularis Injury
- Sacroiliac Joint Injury

Radiography

- Standing lateral radiographs are the preferred method of evaluating slippage of the vertebrae in persons with spondylolisthesis, and they are an excellent means of monitoring for progression of the condition.

- The standing lateral view is best because the translation occurs in the sagittal plane and is often accentuated during standing (due to the oblique orientation of the lower lumbosacral intervertebral disc spaces).

- Standing flexion/extension films should be obtained to assess the degree of instability of the involved vertebrae. These radiographs are also useful in detecting an occult spondylolisthesis.

- The anteroposterior view offers limited information in mild cases of spondylolisthesis; however, in cases of severe slips, this view may reveal the so-called reverse Napoleon hat sign because the L5 vertebra is viewed end-on through the sacrum, giving rise to the appearance of an upside-down Napoleon hat.

- Oblique films are best for evaluating the integrity of the pars interarticularis. A defect is seen as a collar on the neck of the Scotty dog.

Figure 64 Spondylolysis showing 'scotty dog'
Grading spondylolisthesis
Meyerding technique: This involves dividing the superior aspect of the vertebra below the slip into 4 equal divisions, as is observed on a lateral radiograph. Assess where the posterior arch of the slipped vertebral body lies with respect to these 4 quadrants.

- Grade 1: Less than 25% slippage
- Grade 2: Between 25% and 50% slippage
- Grade 3: Between 50% and 75% slippage
- Grade 4: Between 75% and 100% slippage
- Grade 5: Greater than 100% slippage (also called spondyloptosis)

Lumbosacral Spondylolisthesis Treatment & Management

Acute Phase Physical Therapy

As a general rule, physical therapy should not be started until after an adequate rest period and once pain with daily activities has subsided.

The goals of physical therapy are to decrease extension stresses of the lumbar spine and to strengthen elements that promote an antilordotic posture. This consists of exercises to strengthen the abdominal muscles (e.g., William flexion-type exercises) and flexibility programs to stretch the spinal extensor muscles, hamstrings, and lumbodorsal fascia.

Bracing with a thoracolumbosacral orthosis (e.g., Boston antilordotic brace) may offer relief for those who do not respond to activity restrictions or whose daily activities are producing symptoms. This type of bracing is usually effective in most patients with less than 50% slippage. The brace is generally worn for 3-6 months and may be worn during activity.

If the slippage is less than 50% but the patient is symptomatic, then non-operative therapy (e.g., stretching and strengthening exercises, antilordotic brace and activity modification) is instituted. If pain continues to persist, then a spinal fusion is recommended.

Occupational Therapy

Avoidance of heavy-duty labour or activities with repetitive lumbar extension is necessary to allow healing to occur. An occupational therapist can be very beneficial for those individuals who need instructions and compensatory strategies for activities of daily living.

Recreational Therapy

Restriction from sports and other activities that require repetitive hyperextension may be sufficient treatment in young athletes. Patients with grade 2 slippage are generally instructed to avoid hyperextension loading of the spine after symptoms resolve with conservative treatment.

Medical Issues/Complications

Younger patients require more careful observation, even if the initial symptoms resolve, because of their greater risk for progression. In an asymptomatic child with slippage up to 25% (grade 1), initially observe with radiographs every 4-6 months if younger than age 10 years, semi-annually until age 15 years, then annually until the end of growth. No limitation
of activities is required, but the patient is advised to avoid occupations that entail heavy labour. If the slippage is 26-50% (grade 2) and the patient is asymptomatic, then the treatment is the same as for the grade 1 slippage but with a warning against participation in contact sports or sports requiring lumbar hyperextension (e.g., football, gymnastics). In general, the results of conservative management are good in most athletes with Grade I or II slips.

Complications include slip progression, loss of motion segments, neurologic deficit (e.g., cauda equina syndrome, radiculopathy [greatest risk with >50% slippage]), and residual deformity (following fusion of a high-grade spondylolisthesis).

**Surgical Intervention**

Surgery is indicated for skeletally immature patients with greater than 30-50% slippage (with or without symptoms) because they are at greater risk for progression, in the event of progressive neurologic deficit, or in those with pain persisting for more than 6-12 months that has not been relieved with rest and immobilization with any degree of slip. Spondylolysis or low-grade spondylolisthesis may be managed non-operatively.

Options for operative management include direct repair of the spondylolytic defect, fusion in situ, reduction and fusion, and vertebrectomy. Ideally, repair of a pars defect is for young patients with spondylolysis but no spondylolisthesis. Best results are observed in those with a lytic defect between L1 and L4. L5 defects yield less predictable results. Disc degeneration as seen on MRI is a relative contraindication. Slippage of greater than 2 mm decreases the likelihood of successful repair.

Fusion in situ at the involved level is the criterion standard of surgical treatment for most patients in whom conservative management fails. Fusion in situ is recommended for patients with persistent, symptomatic, low-grade spondylolisthesis and for patients who are not candidates for repair of the pars defect. The desire to participate in a contact sport should not be the sole indication for a fusion.

![Figure 65 Spondylolysis fixation](image)

Decompression and fusion are typically performed in cases of dural sac compression with the presence of bowel or bladder dysfunction or significant motor deficits. Decompression is never performed without concomitant fusion. Pedicle screw fixation enables rapid mobilization and early ambulation after decompression and fusion. Fixation may be beneficial in repairing pseudoarthrosis and, in the face of laminectomy, in preventing further slippage while awaiting fusion.
Spondylolisthesis reduction is performed either through closed or open procedures. Reduction serves to correct lumbosacral kyphosis and to diminish sagittal translation observed in high-grade slips. Vertebrectomy may be used to treat spondyloptosis (grade 5 spondylolisthesis), as an alternative procedure to reduction or fusion in situ. The postoperative rate of permanent neurologic deficits is high (25-30%), although many are pre-existent. This does not appear to be balanced by improved results; fusion in situ has achieved similar clinical outcomes with a lower complication rate.

**Spondylolysis**

Spondylolysis is a defect of a vertebra. More specifically it is defined as a defect in the pars interarticularis of the vertebral arch. It can progress until one or more vertebrae slip out of place which is then called spondylolisthesis (see above).

The great majority of cases occur in the lowest of the lumbar vertebrae (L5), but spondylolysis may also occur in the other lumbar vertebrae, as well as in the thoracic vertebrae.

Spondylolysis occurs in three to six percent of the population.

Spondylolysis pain can lead to reduced mobility and inactivity. Inactivity can result in weight gain, loss of bone density, and loss of muscle strength and flexibility of other areas of the body.

It is diagnosed via oblique X-ray (Scotty dog), or CT scan, showing a flaw or damage of the pars interarticularis.

![Figure 66 Spondylolysis on oblique X-Ray](image)

![Figure 67 Spondylolysis on CT scan](image)
Spinal stenosis

Spinal stenosis is an abnormal narrowing (stenosis) of the spinal canal that may occur in any of the regions of the spine. This narrowing causes a restriction to the spinal canal, resulting in a neurological deficit.

Symptoms include:

- Pain
- Numbness
- Paraesthesia
- Loss of motor control

The location of the stenosis determines which area of the body is affected. With spinal stenosis, the spinal canal is narrowed at the vertebral foramen, which is a foramen between the vertebrae where the spinal cord (in the cervical or thoracic spine) or nerve roots (in the lumbar spine) pass through. There are several types of spinal stenosis, with lumbar stenosis and cervical stenosis being the most frequent. While lumbar spinal stenosis is more common, cervical spinal stenosis is more dangerous because it involves compression of the spinal cord whereas the lumbar spinal stenosis involves compression of the cauda equina.

![Figure 68 Spinal Stenosis](image)

Types

The most common forms are cervical spinal stenosis, at the level of the neck, and lumbar spinal stenosis, at the level of the lower back. Thoracic spinal stenosis, at the level of the mid-back, is much less common.

In lumbar stenosis, the spinal nerve roots in the lower back are compressed which can lead to symptoms of sciatica (tingling, weakness, or numbness that radiates from the low back and into the buttocks and legs).

Cervical spinal stenosis can be far more dangerous by compressing the spinal cord. Cervical canal stenosis may lead to serious symptoms such as major body weakness and paralysis. Such severe spinal stenosis symptoms are virtually absent in lumbar stenosis, however, as the spinal cord terminates at the top end of the adult lumbar spine, with only nerve roots (cauda equina) continuing further down. Cervical spinal stenosis is a condition involving narrowing of the spinal canal at the level of the neck. It is frequently due to chronic degeneration, but may also be congenital or traumatic. Treatment frequently is surgical.
What Causes Spinal Stenosis?

There are many potential causes for spinal stenosis, including:

- **Aging:** With age, the body's ligaments (tough connective tissues between the bones in the spine) can thicken. Spurs (small growths) may develop on the bones and into the spinal canal. The cushioning disks between the vertebrae may begin to deteriorate. The facet joints (flat surfaces on each vertebra that form the spinal column) also may begin to break down. All of these factors can cause the spaces in the spine to narrow.

- **Arthritis:** Two forms of arthritis that may affect the spine are osteoarthritis and rheumatoid arthritis.

- **Heredity:** If the spinal canal is too small at birth, symptoms of spinal stenosis may show up in a relatively young person. Structural deformities of the involved vertebrae can cause narrowing of the spinal canal.

- **Instability of the spine, or spondylolisthesis:** When one vertebra slips forward on another, which can narrow the spinal canal.

- **Tumours of the spine:** Abnormal growths of soft tissue may affect the spinal canal directly by causing inflammation or by growth of tissue into the canal. Tissue growth may lead to bone resorption (bone loss due to over-activity of certain bone cells) or displacement of bone and the eventual collapse of the supporting framework of the spinal column.

- **Trauma:** Accidents and injuries may either dislocate the spine and the spinal canal or cause burst fractures that produce fragments of bone that penetrate the canal.

When Should Surgery Be Considered and What Is Involved?

In many cases, the conditions causing spinal stenosis cannot be permanently altered by nonsurgical treatment, even though these measures may relieve pain for a period of time. To determine how much nonsurgical treatment will help, a doctor may recommend such treatment first. However, surgery might be considered immediately if a patient has numbness or weakness that interferes with walking, impaired bowel or bladder function, or other neurological involvement. The effectiveness of nonsurgical treatments, the extent of the patient's pain, and the patient's preferences may all factor into whether or not to have surgery.

The purpose of surgery is to relieve pressure on the spinal cord or nerves and restore and maintain alignment and strength of the spine. This can be done by removing, trimming, or adjusting diseased parts that are causing the pressure or loss of alignment. The most common surgery is called decompressive laminectomy: removal of the lamina (roof) of one or more vertebrae to create more space for the nerves. A surgeon may perform a laminectomy with or without fusing vertebrae or removing part of a disk. Various devices
may be used to enhance fusion and strengthen unstable segments of the spine following decompression surgery.

Patients with spinal stenosis caused by spinal trauma or achondroplasia may need surgery at a young age. When surgery is required in patients with achondroplasia, laminectomy (removal of the roof) without fusion is usually sufficient.

**Facet syndrome**

Facet syndrome is a syndrome in which the zygapophyseal joints cause back pain. In this respect, it may also be known as spondylarthrosis; a condition affecting the facet joints of the spine.

55% of facet syndrome cases occur in cervical vertebrae, and 31% in lumbar. Facet syndrome can progress to spinal osteoarthritis, which is known as spondylosis. Pathology of the C1-C2 (atlantoaxial) joint, the most mobile of all vertebral segments, accounts for 4% of all spondylosis.

It can be generally regarded as a progression of 'wear and tear' of the spine; narrowing of the disc space. If the weight bearing part of the vertebral column, the disc, is narrowed, then the facet joints may begin to bear weight. The problem is that the facet joints are not designed to bear weight and they protest.

It might be said that inflamed facets can cause a powerful muscle spasm. On the other hand, it can be seen that any muscle tightness can cause compression along the axis of the spine, resulting in facet syndrome.

**Intervertebral Disc Herniation**

Spinal disc herniation (commonly called a slipped disc, though this term is not medically accurate as the spinal discs are firmly attached between the vertebrae and cannot "slip") is a medical condition affecting the spine in which a tear in the outer, fibrous ring (*annulus fibrosus*) of an intervertebral disc allows the soft, central portion (*nucleus pulposus*) to bulge out beyond the damaged outer rings.

Disc herniation is usually due to age related degeneration of the annulus fibrosus, although not always so and trauma, lifting injuries, or straining have been implicated. Here, though, it is important to say that we don’t really know as there is not camera during the damage process; we only take photographs ‘afterwards’ to try to explain the pain. It is a working, structural, model that justifies our treatment protocol (structural repair).

Tears are almost always posterolateral in nature owing to the presence of the posterior longitudinal ligament in the spinal canal. This tear in the disc ring may result in the release of inflammatory chemical mediators, which may directly cause severe pain, even in the absence of nerve root compression.

Disc herniations are normally a further development of a previously existing disc "protrusion", a condition in which the outermost layers of the *annulus fibrosus* are still intact, but can bulge when the disc is under pressure. In contrast to a herniation, none of the *nucleus pulposus* escapes beyond the outer layers. Most minor herniations heal within several weeks. Anti-inflammatory treatments for pain associated with disc herniation, protrusion, bulge, or disc tear are generally effective.
Severe herniations may not heal of their own accord and may require surgical intervention.
Lumbosacral transitional vertebrae
The design of the spine is complex enough already, with the majority of the body’s weight being transferred through the L5/S1 junction. Normally there are 5 vertebrae in both the lumbar and sacral regions of the spine, with the bones of S1-5 fused together. To make this more awkward, congenital variation can occur at the L5/S1 level.

![X-ray of lumbosacral region](image)

**Figure 71 X-ray of lumbosacral region**

The picture above demonstrates the principles of lumbarisation and sacralisation.

With all this, though, there are two main possibilities for a lumbosacral transitional vertebra:

- **Lumbarisation of S1**
- **Sacralisation of L5**

These sound like deliberately confusing terms, but remember that the number of bones in each section of the spine remain the same, any changes in this number causes changes that can occur in the function of the region:

- In a lumbarised S1, the S1 vertebra functionally becomes part of the lumbar spine
  - Effectively creating a longer lumbar spine
- In a sacralised L5, the L5 vertebra functionally becomes part of the sacrum
  - Effectively creating a shorter lumbar spine

The **Castellvi classification** is used for lumbosacral transitional vertebra

- **Type I** - enlarged and dysplastic transverse (at least 19 mm)
  - Ia - unilateral
  - Ib - bilateral
- **Type II** - pseudoarticulation of the transverse process and sacrum with incomplete lumbarisation/sacralisation; enlargement of the transverse process with pseudoarthrosis
  - Ila - unilateral
- **Type III** - transverse process fuses with sacrum and there is complete lumbarisation or sacralisation, enlarged transverse process with complete fusion
  - IIIa - unilateral
  - IIIb - bilateral
- **Type IV** - type IIa on one side and type III on contralateral side

![Figure 72 Castellvi classification](image)
**Lumbarisation of S1**

This congenital situation can cause confusion, even to the experienced eye. This is one of those confusing transitional vertebra cases that defies a clear categorisation (left).

There are 6 vertebra that demonstrate lumbar characteristics. The vertebra above the vertebra annotated with "1" has associated ribs. Are these the 12th ribs or are these the 11th ribs with congenital absence of the 12th ribs?

Seeing this picture, and recalling that the sacroiliac joints (see inset) extend between S1-3, questions arise about whether this condition can be blamed on low back pain when it occurs. This X-ray clearly shows that the S/I joints are of reasonably normal configuration, but the bony apparatus does not extend to include S1. It is sometimes referred to as ‘L6’, but this is not strictly true as there is no L6 nerve root.

**Figure 74 Sacralised L5 - A/P view**

"Lumbarized S1"

- note the widened transverse processes
- (incidentally, this lumbarosmal variation classifies as a Castellani B, which a recent study claimed occurs in 8.5% of the population).
- (mild spina bifida at S2)
The association of lumbosacral transitional vertebra and low back pain is known as Bertolotti’s syndrome. This syndrome was characterized by the presence of a variation of the fifth lumbar vertebra having a large transverse process, either articulated or fused with sacral base or iliac crest and producing a chronic, persistent low back pain due to arthritic changes occurring at the site of pseudoarthrosis.

It may be a sweeping statement, but it might be said that such a condition only becomes a problem if the person has overt symptoms. Lumbarisation of S1 can potentially lead to instability, especially if associated with a pars interarticularis defect, leading to a spondylolisthesis, as seen here.

Sacralisation of L5
Sacralisation of L5 is the other variant of lumbosacral transitional vertebrae

Lumbosacral region is one such region there are so many puzzling stresses and strains, renders the question more than ordinary interest. Occult sacralisation has been described by O’Connell 1951, where the sacrum is high in the pelvis and the spinous process of the last lumbar vertebra may be with or just below the iliac crests. The lumbar spine shows five vertebrae, but the sacrum may be composed of six vertebral segments. As these are not cases of frank sacralisation of fifth lumbar vertebra, the term “Occult Sacralisation” has been used to describe the abnormality.
Figure 77 Sacralisation of L5

The upper picture demonstrates a type iv, according to Castellvi, whereas on the left is a type iiA.

Both will significantly reduce any motion between the two bones.

With the two examples, the types iiA may permit some movement on the left (the right demonstrating at least a pseudoarthrosis on the right, but the type iv, as it is fused bilaterally, will allow no movement at all.

This type of condition reduces the number of bones in the lumbar spine, possibly reducing the overall movement, but also possibly putting a greater demand upon the lumbar structures and joints that are remaining.

Figure 78 Sacralisation of L5 on CT scan

Disc bulge / herniation occurs nearly nine times more commonly at the interspace immediately above the lumbar transitional vertebra than at any other level.

Lumbosacral transitional vertebra increases the risk of early degeneration in the upper disc. The presence of a transitional vertebra should be noticed when morphologic methods are used in research on lumbosacral spine.
Compression Fractures of the Spine

The vertebral bodies and processes consist of compact bones surrounding trabecular bone. In cases of osteoporosis and/or trauma, fractures can occur. The vertebral bodies in the lower thoracic and upper lumbar spine have mainly parallel superior and inferior epiphyseal plates.

The type of fracture in the spine that is typically caused by osteoporosis is generally referred to as a compression fracture.

- A compression fracture is usually defined as a vertebral bone in the spine that has decreased at least 15 to 20% in height due to fracture.
- These compression fractures can occur in vertebrae anywhere in the spine, but they tend to occur most commonly in the upper back (thoracic spine), particularly in the lower vertebrae of that section of the spine (e.g. T10, T11, T12).
- They rarely occur above the T7 level of the spine, occurring in the upper lumbar segments as well, such as L1.

Classification of Fractures

Classifying the various types of spinal fractures can be confusing because there are several classification methods. Here the focus will be on the most basic and the most generally accepted classifications for spinal fractures.

Denis Classification: The Three-column Concept

The three-column spine was first introduced by Dr. Francis Denis in his aptly named paper, "The Three Column Spine and its Significance in the Classification of Acute Thoracolumbar..."
Spinal Injuries”. This paper, published in 1983, proposed a new biomechanical model for spinal stability that challenged Dr. Frank Holdsworth’s two column model from the 1960s.

Denis’ three column model proposes that the thoracolumbar spine can be divided into three columns. The first column includes the anterior longitudinal ligament (ALL) up to the first half of the bony vertebral body. The second column includes the second half (i.e.: more posterior half) of the vertebral body, up to, and including the posterior longitudinal ligament (PLL). The third column includes the pedicles, spinal cord/thecal sac, lamina, transverse processes, facet joints, spinous process, and the posterior ligaments (i.e. supraspinous, interspinous, and ligamentum flavum).

The purpose of Denis’ model was to delineate which injuries to the thoracolumbar spine were considered "unstable". This delineation was important because it determined which patient's required operative treatment of their spine.

Simply stated, an unstable spine was present if two or more of the columns were involved in the injury. However, it is important to note that every rule can be broken, so not all injuries to the thoracolumbar spine follow this rule. Part of the "art" of practicing spine surgery is determining which two-column injuries can be left alone and managed non-operatively, and which truly require operative intervention.

Based on Denis' model he classified specific types of injuries. Injuries to the anterior column only were called "compression fractures". Damage to the anterior and middle columns were known as "burst fractures". Injury to the middle and posterior column were known as "flexion-distraction" injuries, or more colloquially as "seat-belt type" injuries. Finally, damage to all three columns were classified as "fracture-dislocation" injuries.

<table>
<thead>
<tr>
<th>Classification of Thoracolumbar Injuries Using the Three Column Model</th>
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<td>Columns</td>
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<tr>
<td>Anterior</td>
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<td>Anterior and Middle</td>
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<td>Middle and Posterior</td>
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<td>Anterior, middle, and posterior</td>
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This system divides the spine into three columns (when viewing the spine from the side):

- **Anterior Column**: This is the front part of the vertebra—the part that faces in towards your body. The anterior column is the front half of the vertebral body and intervertebral disc. There's a ligament on the front of the spine called the anterior longitudinal ligament; that's part of the anterior column as well.

- **Middle Column**: This is the key part of spinal stability. It's the back half of the vertebral body and intervertebral disc. There's a ligament on the back of the vertebral body called the posterior longitudinal ligament; that's part of the middle column as well. If there's a fracture in the middle column in addition to the anterior or posterior column, you're much more likely to have nerve damage and spinal instability. As long as the middle column stays intact, you have a better chance of having a stable fracture.

- **Posterior Column**: All the parts of the vertebra that are on the back side make up the posterior column. This includes the pedicles, lamina, facet joints, and spinous process (that's the bony part you can feel when you run your finger up your spine).

The three-column concept makes it easier to visualize spinal fractures. It also makes it easier to understand why some fractures are more stable than others. For example, if you have a fracture just in the anterior column, the spine should be able to still carry your weight well enough.

However, if you fracture the anterior and middle column, it's more likely that your spine will be unstable. Since the middle column connects the anterior and posterior columns, if that part is fractured, it will be harder for the spine to function well.
In very simple terms, unstable fractures require definitive treatment. Treatment for unstable spine injuries is usually operative, although rigid immobilization with bracing is sometimes used in certain circumstances.

The treatment of each fracture type is beyond the scope of this article (and discussed in more detail elsewhere on this site), but suffice it to stay that for the most part (and again, every rule was made to be broken) two or three column injures = unstable = surgery.

**Major and Minor Fractures**

This is the simplest way to talk about fractures: major or minor.

A minor fracture means that a part of the posterior (back side) elements of the vertebra has broken—the parts that aren't as vital to spinal stability. The posterior elements include the spinous process and the facet joints (also called the articular processes). If you fracture this part of the vertebrae, it's usually not too serious.

A major fracture means that part of the vertebral body, the pedicles, or the lamina has fractured. Fracturing the vertebral body is considered major because it helps carry so much weight and distribute the force of your movements. If it's broken, you can have serious problems with the vertebrae lining up correctly. Fracturing the pedicles or lamina is dangerous because of the increased possibility of nerve damage. Additionally, the pedicles and lamina provide a lot of necessary support to keep your spine stable. If they fracture, your spine may be unstable.

**Stable and Unstable Fractures**

Stable and unstable is another basic classification for spinal fractures.

Stable fractures don't cause spinal deformity or neurologic (nerve) problems. With a stable fracture, the spine can still carry and distribute your weight pretty well (not as well as if there weren't a fracture, but it's still able to function with a stable fracture).
Unstable fractures make it difficult for the spine to carry and distribute weight. Unstable fractures have a chance of progressing and causing further damage. They may also cause spinal deformity.

**Specific Fracture Types**
Beyond whether a fracture is stable or unstable, major or minor, you should understand the different ways a vertebra can break. As your doctor describes your fracture, you may hear terms such as:

- **Compression Fracture:** This type of fracture is very common in patients with osteoporosis, or patients whose bones have been weakened by other diseases (such as bone cancer). The vertebra can absorb so much pressure; if there's a sudden force of a lot of pressure, the bone may not be able to handle the stress. The vertebra can fracture then.
  - A wedge fracture is a subtype of compression fracture. With a wedge fracture, part of the vertebra—usually the anterior (front) part—collapses under pressure and becomes wedge shaped.

- **Burst Fracture:** Burst fractures are caused by severe trauma (e.g., car accident). They happen when the vertebra is essentially crushed by extreme forces. Unlike compression fractures, it's not just one part of the vertebra that's fractured. In a burst fracture, the vertebra is fractured in multiple places. Because the vertebra is crushed completely, bony fragments can spread out and cause spinal cord injury. Burst fractures are more severe than compression fractures.

- **Flexion-distraction Fractures:** If you're in a car accident where your body is pushed forward, you may get a flexion-distraction fracture. Your spine is made to flex forward, but if there's a sudden forward movement that places incredible stress on the spine, it may break a vertebra or vertebrae. Thinking of the three-column concept, a flexion-distraction fracture usually has fractures in the posterior and middle column.

- **Fracture-dislocation:** If you have any of the above fractures and the vertebra(e) moves significantly (dislocation), you have a fracture-dislocation. Usually, these fractures involve all three columns from the three-column concept, and they make your spine very unstable.
The term "wedge fracture" is used because the fracture usually occurs in the front of the vertebra, collapsing the bone in the front of the spine and leaving the back of the same bone unchanged. This process results in a wedge-shaped vertebra. A wedge compression fracture is generally a mechanically stable fracture pattern.

A wedge fracture occurs when the anterior of the vertebral body is crushed in relation to the posterior, as seen here. It may result in a flexion deformity of the spine.

If a wedge fracture causes a flexion deformity, it can be assessed by drawing lines along the posterior vertebral bodies (PVB). A line drawn along these should be continuous. A break suggests a translational deformity, as seen here in this lateral lumbar X-ray.
While wedge fractures are the most common type of compression fracture, there are other types as well, such as:

- **Crush fracture** - If the entire bone breaks, rather than just the front of the vertebra, it may be called a crush fracture.

  ![Figure 85 Crush fracture](image)

Depending upon the severity, this may result in a reduction in the length of the spine.

- **Burst fracture** - This type of fracture involves some loss of the height in both the front and back walls of the vertebral body (rather than just the front of the vertebra). It may even cause detached fragments, as seen here in the anterior part of the vertebral body. Making this distinction is important because burst fractures can be unstable and result in progressive deformity or neurologic compromise.

  ![Figure 86 Burst Fracture](image)

As these X-rays demonstrate, they are easily seen on lateral X-ray. They can also be seen on A/P X-ray as well.

![Figure 87 Crush fracture L3](image)

This X-ray is marked with arrows showing the positions of the pedicles in the lumbar spine. The crush fracture has occurred at the level of L3, with the pedicles being pushed outwards, bilaterally. This arrangement fails to follow the
normal arrangement of the pedicles gradually widening from top to bottom.

**Compression Fracture Symptoms**
Vertebral fractures are usually followed by acute back pain, and may lead to chronic pain, deformity (thoracic kyphosis, commonly referred to as a dowager's hump), loss of height, crowding of internal organs, and loss of muscle and aerobic conditioning due to lack of activity and exercise. A combination of the above problems from vertebral fractures can also lead to changes in the individual's self-image, which in turn can adversely affect self-esteem and ability to carry on the activities of daily living.

Because the majority of damage is limited to the front of the vertebral column, the fracture is usually stable and rarely associated with any nerve or spinal cord damage.

![Figure 88 Crush fracture with spinal cord damage](image-url)

This MRI scan shows a crush fracture. It also demonstrates a fusiform swelling of the spinal cord at that level, suggesting substantial swelling there. It is here that the MRI scan is very useful, as X-ray or CT examination would not show this.

**Fracture-Dislocations**
Fracture-dislocations of the thoracolumbar spine are highly unstable injuries. Although they may display various combinations of damage to both the anterior and posterior elements, the unique injury feature is translational deformity, which can occur in the sagittal and/or coronal planes.

![Figure 89 Flexion traction injury](image-url)

Type A and B occur at one level
Type C and D occur at two levels
Type A are bony one-level injuries
Type B are one-level ligamentous injuries
Type C injuries are two-level injuries that occur through bone and/or ligament.
Any and all of these are likely to cause spinal cord damage.

Canal compromise rarely occurs from displaced fractures of the posterior elements. In the axial CT scan here, a large piece of fractured lamina intruded on the spinal canal of this patient who had sustained a high-energy injury to the thoracic spine.

Figure 91 Fragment of bone in spinal canal

Treatment
A careful review of the data suggests that the nonsurgical management of stable thoracolumbar burst fractures (defined as fractures that have no PLC injury without neurologic deficit) can provide good results. Although many investigators have suggested the use of various x-ray measurement criteria for non-operative treatment such as less than 25 degrees to 30 degrees of kyphosis, less than 50% height loss, the absence of interspinous process widening, and less than 50% canal compromise, we believe that MRI evidence of discontinuity or continuity of the posterior ligamentous complex (PLC) is very important.

Provided the patient is neurologically intact and the PLC is intact, greater amounts of canal compromise or height loss can be accepted. Doctors prefer to keep the patient flat and on log-roll precautions until a thoracolumbar sacral orthosis (TLSO) [brace] is in place. The patient then undergoes a trial of standing x-rays before ambulation is permitted. X-ray and clinical follow-up examinations are scheduled at 2 weeks, 1 month, 2 months, and 3 months. At the 3-month follow-up, x-rays are made out of the brace to ensure stable alignment.

**Surgical Treatment**

Anterior and posterior approaches have been advocated for decompression and stabilization; both having advantages and disadvantages. Although there is uncertainty regarding the optimal surgical treatment of thoracolumbar burst fractures, so fixation is favoured in cases of neurological compression of the spinal cord and increasing kyphotic deformity. In addition, fractures in multiple injured patients or those in whom bracing cannot be effectively employed, due to other injuries or body habitus, may benefit from internal stabilization.

**Posterior Surgery**

Advocates of posterior surgery cite various advantages compared to anterior surgery for thoracolumbar burst fractures. First, it avoids the morbidity of anterior exposure in patients who potentially have concomitant pulmonary or abdominal injuries and shorter operative times and decreased blood loss, and functional outcomes are similar to those following anterior surgery.

*Figure 92 Posterior spinal fixation*

Here, special screws are inserted through the axis of the pedicles, into the bodies of the vertebrae; at and either side of the damaged vertebra. Then rods are attached and secured,
fixing the vertebrae together. The role of posterior surgery for burst fractures is primarily for realignment and stabilization

![Posterior Spinal Fixation X-ray](image)

The X-ray here demonstrates such posterior fixation of the spine. Posterior fixation alone cannot reconstitute anterior column support, however, and is therefore somewhat weaker in compression than anterior fixation. This has led to a higher incidence of progressive kyphosis and instrumentation failure when treating highly comminuted fractures

**Anterior Surgery**

Anterior surgery for thoracolumbar burst fractures is primarily indicated for decompression of the neural elements.

![Anterior Spinal Fixation with Kaneda Device](image)

Note that a titanium mesh cage (filled with salvaged fractured bone and supplemented by allograft) was used to reconstruct the anterior column. If an anterior corporectomy (removal of vertebral body) is undertaken to decompress the spine, it must be understood that the spine is always destabilized. To remedy this situation the vertebral body defect must be replaced with a supportive strut that will also result in bony fusion. A structural, bony, autograft can be harvested from rib, fibula, or the iliac crest and inserted.
Another method of repairing crush fractures of the spine, if the alignment or length is compromised, or if there is neurological deficit. There is a technique for injection of bone cement into the body of the vertebra itself. The technique is called kyphoplasty.

The series of diagrams demonstrate that a hole is drilled into the vertebra, through the pedicle (the same route as the screw insertion of spinal fixation). Then a balloon is inserted and inflated (similar to angioplasty with the insertion of a stent), then the bone cement is injected. This technique helps maintain the structure and width of the vertebral body and help minimise any kyphosis resultant of the crush fracture.
Whiplash and Cervical Spine Injuries
The majority of road traffic accidents precipitate a reactive spasm and chronic painful tension in the cervical spine and shoulders. This possibly associated with a mild, non-inflammatory, sprain of the associated ligaments. The bulk of these can be treated with soft tissue and myofascial techniques. There will always be the incident of severe injury.

This review is based on a presentation given by Adam Flanders and adapted for the Radiology Assistant by Robin Smithuis.

- Approximately 3% of patients who present to the emergency department as the result of a motor vehicle accident or fall have a major injury to the cervical spine. 10-20% patients with head injury also have a cervical spine injury.
- Up to 17% of patients have a missed or delayed diagnosis of cervical spine injury, with a risk of permanent neurologic deficit after missed injury of 29%.
- Most cervical spine fractures occur predominantly at two levels.
- One third of injuries occur at the level of C2, and one half of injuries occur at the level of C6 or C7.

In this overview we will discuss the most common cervical spine injuries.

Flexion injuries

The most common fracture mechanism in cervical injuries is hyperflexion.

- **Anterior subluxation** occurs when rupture of the posterior ligamentous complex (PCL). Since the anterior and middle columns remain intact, this fracture is stable.
- **Simple wedge fracture** is the result of a pure flexion injury. The PCL remain intact. Anterior wedging of 3mm or more suggests fracture. Increased concavity along with increased density due to bony impaction. Usually involves the upper endplate.
- **Unstable wedge fracture** is an unstable flexion injury due to damage to both the anterior column (anterior wedge fracture) as the posterior column (interspinous ligament).
- **Unilateral interfacet dislocation** is due to both flexion and rotation.
- **Bilateral interfacet dislocation** is the result of extreme flexion. BID is unstable and is associated with a high incidence of cord damage.
- **Flexion teardrop fracture** is the result of extreme flexion with axial loading. It is unstable and is associated with a high incidence of cord damage.
Anterior atlantoaxial dislocation

**Figure 96 Cervical flexion injuries**

**Extension injuries**

- **Hangman’s fracture**
  - Traumatic spondylolisthesis and fracture of C2
- **Extension teardrop fracture**
  - Avulsion fracture via ALL on anterior body of C3
- **Hyperextension in pre-existing spondylosis**
  - ‘Open mouth fracture’
  - Disruption of anterior ligaments and intervertebral disc (here) at C3/4
Axial compression injuries

- **Jefferson fracture** is a burst fracture of the ring of C1 with lateral displacement of both articular masses
- Burst fracture at lower cervical level

Figure 97 Cervical extension injuries

Figure 98 Jefferson fracture of C1
Stability

With bony damage and/or disruption of ligaments, stability at the level of injury may be compromised, possibly with consequent spinal cord injury

Unstable fractures:

- **Flexion**
  - Bilateral interfacetal dislocation
  - Flexion teardrop fracture
  - Wedge fracture with posterior ligamentous rupture
- **Extension**
  - Odontoid fracture type II
  - Hangman's fracture
  - Extension teardrop fracture
- **Vertical compression**
  - Burst fracture, e.g. Jefferson fracture

Spinal cord injury

There are two types of injury to the spinal cord:

- Non-haemorrhagic with only high signal on MRI due to oedema.
- Haemorrhagic with areas of low signal intensity within the area of oedema.
Here, there is a strong correlation between the duration of the spinal cord oedema and the clinical outcome. The most important factor however is whether there is haemorrhage, since haemorrhagic spinal cord injury has an extremely poor outcome.

The chart below is showing the motor recovery rate for patients with oedema alone (in blue) versus oedema plus cord haemorrhage (in red). The motor recovery rate is for the legs only.

![Motor Recovery Chart]

**Figure 100 Incidence of haemorrhagic spinal cord injury**

**Spinal cord syndromes:**

1. **Central cord syndrome**
   - Most common incomplete cord syndrome.
   - Frequently found in elderly with underlying spondylosis or younger people with severe extension injury (figure).
   - Upper extremity deficit is greater than lower extremity deficit, because the lower extremity corticospinal tracts are located lateral in the cord.

2. **Anterior cord syndrome**
   - Seen in flexion injuries e.g. burst fracture, flexion tear drop fracture and herniated disk.
   - Presents with immediate paralysis, because the corticospinal tracts are located in the anterior aspect of the spinal cord.

3. **Brown-Sequard syndrome**
   - Ipsilateral motor weakness and contralateral sensory deficit due to hemisection of the spinal cord.
   - Brown-Sequard syndrome may result from rotational injury such as fracture-dislocation or from penetrating trauma such as stab wound.

4. **Posterior cord syndrome**
   - Uncommon syndrome due to extension injury.
   - Loss of positioning sense due to disruption of dorsal columns.
   - Good prognosis.

5. **Complete spinal cord injury**
   - Total absence of sensation and motor function caudal to the level of injury.
Hyperflexion sprain injuries are injuries to the soft tissues of the spine without fracture. X-rays of this can only suspect damage when there is angulation or translation, but MRI scans will demonstrate subtle injuries to the soft tissues.

On the left images of a patient who has been in a car accident and complained of neck pain. The x-rays were normal and there were no neurological symptoms.
The findings are:

- Oedema in the posterior soft tissues indicating a hyperflexion injury
- Oedema in the vertebrae of the lower C-spine and upper T-spine indicating bone bruise as a result of axial loading.

In this patient we can conclude that there was mild hyperflexion strain and we do not know if a special treatment is required, since these were isolated MR-findings without evidence of fracture or abnormal positioning. However, there is controversy regarding the meaning of soft tissue abnormalities detected only on MRI.

- Signal changes do not necessarily equate with structural failure.
- These findings still require better validation.
- In trauma centres up to 25% of all patients with neck injury have signal abnormalities on MRI and the significance is indeterminate.

Below are the images of a 44-year-old female, who sustained a fall on the ice. She subsequently had a second fall the following morning, where after she had complete loss of motor and sensation. On physical examination, there was lower extremity paraparesis with some upper extremity weakness on the right. Central cord injury was proposed initially. The radiographs were normal.

![Figure 103 Cervical hyperflexion injury in 44 year old](image)

The findings are:

- Small bone fragments coming off the superior and inferior facets
- Widened interspinous space at C5-6
- Soft tissue swelling at this level posteriorly
- Subtle narrowing of the disc space at the C5-6-level
These CT-findings are very subtle and do not seem to match the neurological problem. In such a case MRI is the next step.

![Cervical MRI scan](image)

The MRI-findings are:

- Severe soft tissue injury of the posterior paraspinal structures, especially at the C5-6 level, where the interspinous ligament and the ligamentum flavum is ruptured
- Disruption of the C5-6 disc with migration behind C5
- Large amount of spinal cord oedema

This type of cervical injury can also compromise the vertebral arteries and their blood supply for the brain stem and arterial circle of Willis. It is here that cervical manipulation is contra-indicated.

**Unilateral interfacet dislocation**

Unilateral interfacet dislocation is due to a hyperflexion injury with rotation. The superior facet on one side slides over the inferior facet and becomes locked. This results in an anterior subluxation of the upper vertebral body of about 25% of the A/P diameter of the body. Simple unilateral facet dislocation is a stable injury. 30% of patients have an associated neurologic defect. MRI plays an important role in the diagnosis in order to see if there is disc extrusion leading to cord compression.

![Cervical unilateral dislocation](image)
The next example is a 20-year-old male who had a rollover motor vehicle accident.

The radiographic findings are:

- Hyperflexion at the level of C4/5 with widening of the interspinous space
- Subluxation at the level of C4/5 with about 25% translation (i.e. anteroposition of 25% of the AP diameter of the vertebral body)
- Malalignment of the spinous processes as seen on the A/P-view, which can only be produced by a rotatory injury. The involved spinous process points to the involved side
- Due to the rotation the spinous processes of C4 and C5 seem shorter on the lateral view

The CT confirms the unilateral dislocation. The contralateral facet joint is only distracted.
The MRI-findings are:

![MRI Image with Arrows](image)

- Spinal cord lesion, which can be described as contusion, oedema or non-haemorrhagic spinal cord injury.
- Dislocation of C4/5 facet (yellow arrow)
- Rupture of the spinous ligaments (blue arrow)
- Rupture of the ligamentum flavum.
- Rupture of the disc with migration of disc material on the posterior side of C4 and even on the anterior side of C5 (red arrow)
  The disc space is always disrupted in this kind of injury due to the extreme rotation.

**Bilateral Interfacet Dislocation**

Bilateral interfacet dislocation (BID) is the result of extreme hyperflexion. There is anterior dislocation of the articular masses with disruption of the posterior ligament complex, posterior longitudinal ligament, the disc and usually also the anterior longitudinal ligament.

When the dislocation is complete, the dislocated vertebra is anteriorly displaced one-half of the AP diameter of the vertebral body.

Because of its extensive soft tissue damage and dislocated facet joints, BID is unstable and is associated with a high incidence of cord damage.

![Diagram of BID](image)
The findings are:

- Bilateral interfacet dislocation.
- 50% anteroposition C5/6 as a result of the dislocation.
  - In unilateral dislocation, the anteroposition is usually only 25%.
- Widened space between spinous processes C5 and C6 due to ligament rupture.
- Ruptured disc space.

The MRI findings for the same patient are:

- Soft tissue swelling anteriorly
- Disruption of the disc
- Non-haemorrhagic cord injury
Correction of this was performed via a traction technique in order to regain normal alignment. Progressive weights are used to lengthen the spine until reduction is achieved. In this case, with 60 pounds the facets start to move, but it finally takes about 110 pounds before the neck is reduced. Because someone is holding on to the neck while more weight is added, an actual 'clunk' can be felt in the neck indicating that reduction is achieved.

Below are images of a 15-year old, who was injured during wrestling.

- There is 50% anteroposition of C3 on C4 as a result of bilateral interfacetal dislocation.
- There is complete disruption of the posterior complex.
- This boy had severe neurologic deficit.

![Figure 112 Cervical bilateral dislocation of 15 year old](image)

**Flexion tear drop fracture**

This fracture is the result of a combination of *flexion* and *compression*, which is usually the result of a motor vehicle accident.

The teardrop fragment comes from the anteroinferior aspect of the vertebral body. The larger posterior part of the vertebral body is displaced backward into the spinal canal.

![Figure 113 Flexion teardrop fracture](image)

On X-rays the facet joints and interspinous distances are usually widened and the disk space may be narrowed. 70% of patients have neurologic deficit.
It is an unstable fracture associated with complete disruption of ligaments and anterior cord syndrome.

Below are images of a 21-year-old male who sustained a diving injury, striking his head in a swimming pool. He had immediate onset of upper and lower extremity weakness.

Some would just call this a severe hyperflexion injury, but this entity is better known as a ‘flexion tear drop’ fracture

The findings are:

- Fracture of the body of C5 with a small fragment anteriorly
- Fracture of the spinous processes of C4
- Acute angulation at the level of C5C6 with displacement of C5 in posterior direction

![Figure 114 X-ray tear drop fracture](image)

Additional findings on the CT-images:

- Abnormal positioning of some of the facet joints due to distraction but no dislocation
- Additional fracture of the body of C4 (blue arrow)
- The vertical orientation of the fractures of the bodies of C4 and C5 indicate that there was severe axial loading (red arrow)
- In fact these vertebral bodies kind of ‘exploded’ with propulsion of a bone fragment anteriorly (teardrop) and the larger part posteriorly against the spinal cord.
The MRI findings are:

- Soft tissue injuries anteriorly and posteriorly with flavum and interspinous ligament rupture and CSF leakage.
- Haemorrhagic spinal cord injury!

Hyperextension Injuries
Hangman’s fracture

The Hangman’s fracture is the most common cervical spine fracture. Classically it is an extension-fracture as the hangman puts the knot under the chin to produce maximal extension-force. That is why the hangman’s fracture is discussed in hyperextension injuries. In some situations however it can also be the result of extreme flexion.
The hangman’s fracture is common in diving accidents. Although considered an unstable fracture, it seldom is associated with spinal injury, since the anteroposterior diameter of the spinal canal is greatest at this level, and the fractured pedicles allow decompression. When associated with unilateral or bilateral facet dislocation at the level of C2, this type of hangman’s fracture is unstable and has a high rate of neurologic complications.

Classification of Hangman’s fractures

- **Type I** (65%)
  - hair-line fracture
  - C2-3 disc normal
- **Type II** (28%)
  - displaced C2
• disrupted C2-3 disc
• ligamentous rupture with instability
• C3 anterosuperior compression fracture

• **Type III** (7%)
  • displaced C2
  • C2-3 Bilateral interfacet dislocation
  • Severe instability

Below are images of a restrained passenger in a vehicle going about 55 miles per hour. She ran into a tree at about 9 p.m. the previous night with questionable loss of consciousness. She had cervical tenderness to palpation, but was alert and had no neurologic abnormalities on examination.

![Image of a restrained passenger in a vehicle going about 55 miles per hour.](image)

**Figure 118** Hangman’s fracture following road accident

The findings are:

- Subtle lucent line at the back of the corpus of C2 as seen on the lateral view (arrow).
- Subtle discontinuity of the arch of C2

The CT-images confirm the fracture-lines of the hangman's fracture. They run through the pars interarticularis resulting in a traumatic spondylolysis. In this case there was no neurologic deficit, because the spinal canal is widened at the level of the fracture.
The images here are of a 90-year-old male who tripped and fell on his back and the back of his head. He had immediate quadriparesis after the event with no loss of consciousness.

The (above) X-ray findings are:

- Widening of the disc space C5/6 in the front and narrowing in the back.
- This is also called 'an open book'.
- It tells us that there was a hyperextension injury.
The CT scan (left) shows a hyperextension injury. The small black dots in the disc space are the result of a vacuum phenomenon. The negative pressure resulted in a vacuum phenomenon in the injured disc space.

There is also some hyperdensity at the back of C5/6, which could be a herniated disc or just pre-existing disc degeneration.

In such a patient with spondylosis which has led to narrowing of the canal, a low velocity injury can lead to spinal cord injury.

Figure 122 Cervical open book CT scan

The MRI, below, shows a subtle increase in signal intensity of the spinal cord. Most of the time these patients get a central cord injury. There is only injury to the central part of the cord and these patients have disproportioned weakness of their arms and normal strength in their legs. These injuries can be devastating, although it is uncommon that they are haemorrhagic.

Figure 123 Cervical open book MRI
Extension teardrop fracture
As with flexion teardrop fracture, extension teardrop fracture also manifests with a displaced anteroinferior bony fragment. This fracture occurs when the anterior longitudinal ligament by creating an avulsion fracture by pulling a bony fragment away from the inferior aspect of the vertebra because of the sudden hyperextension. The fragment is a true avulsion, in contrast to the flexion teardrop fracture in which the fragment is produced by compression. This type of fracture is commonly seen in diving accidents and tends to occur at lower cervical levels. It also may be associated with the central cord syndrome due to buckling of the ligamenta flava into spinal canal during the hyperextension phase of injury. This injury is stable in flexion but highly unstable in extension.

Figure 124 Extension teardrop fracture

On the left images of a 70 year old female who fell down ten steps striking her head resulting in a subgaleal hematoma with possible loss of consciousness. There was no neurologic deficit. Notice the anteroinferior bony fragment of C2.

The MRI scan, below, also confirms that this is not a flexion injury, since the soft tissue injury is located anteriorly.

Figure 126 Extension teardrop MRI
Figure 127 Acute cervical spine injury resume
Coccydynia

Coccydynia is a medical term meaning pain in the coccyx or tailbone area. Coccydynia is also known as coccygodynia, coccygeal pain, coccyx pain, or coccalgia.

Figure 128 Coccygeal pain

Structure

Coccydynia occurs in the lowest part of the spine, the coccyx, which represents a vestigial tail, or in other words the “tail bone”. The name coccyx is derived from the Greek word for cuckoo due to its beak like appearance. The coccyx itself is made up of 3 to 5 fused vertebrae. The ventral side of the coccyx is slightly concave whereas the dorsal aspect is slightly convex. Both of these sides have transverse grooves that show where the vestigial coccygeal units had previously fused. The coccyx attaches the sacrum, with the attachment being either a symphysis or synchondrosis, and also to the gluteus maximus muscle, the coccygeal muscle, and the anococcygeal ligament.

Causative factors could be local trauma, like falling on it, but this is not always the case.

Activities that put pressure on the affected area are bicycling, horseback riding, and other activities such as increased sitting that put direct stress on the coccyx. The medical condition is often characterized by pain that worsens with constipation and may be relieved with bowel movement. Rarely, even sexual intercourse can aggravate symptoms.

Treatment

This can take the form of local steroid injection, to relieve symptoms. If it is caused by local trauma, there may be local spasm, damage or fibrosis of the local musculature and this can be treated by physical therapy. It can also be treated by ‘balancing’ the perineal muscles (those of the pelvic diaphragm) by a pelvic diaphragm release (as in craniosacral therapy) or functionally releasing the coccyx through holding it directly (one finger inside the anal canal and the out outside).

Figure 129 Coccygeal correction
Referred symptoms
Referred symptoms is pain perceived at a location other than the site of the painful stimulus. An example is the case of ischemia brought on by a myocardial infarction (heart attack), where pain is often felt in the neck, shoulders, and back rather than in the chest, the site of the injury. The International Association for the Study of Pain, as of 2001, has not officially defined the term; hence several authors have defined the term differently.

Radiation is different from referred pain. The pain related to a myocardial infarction could either be referred pain or pain radiating from the chest. Classically the pain associated with a myocardial infarction is located in the mid or left side of the chest where the heart is actually located. The pain can radiate to the left side of the jaw and into the left arm. Referred pain is when the pain is located away from or adjacent to the organ involved. Referred pain would be when a person has pain only in their jaw or left arm, but not in the chest. Myocardial infarction can rarely present as referred pain and this usually occurs in people with diabetes or older age.

Physicians and scientists have known about referred pain since the late 1880s. Despite an increasing amount of literature on the subject, the mechanism of referred pain is unknown, although there are several hypotheses.

Characteristics of referral
- The size of referred pain is related to the intensity and duration of ongoing/evoked pain.
- Temporal summation is a potent mechanism for generation of referred muscle pain.
- Central hyper-excitability is important for the extent of referred pain.
- Patients with chronic musculoskeletal pains have enlarged referred pain areas to experimental stimuli. The proximal spread of referred muscle pain is seen in patients with chronic musculoskeletal pain and very seldom is it seen in healthy individuals.
- Modality-specific somatosensory changes occur in referred areas, which emphasize the importance of using a multimodal sensory test regime for assessment.
- Referred pain can be difficult to appreciate as they have origins via ‘shared’ neurological origins, or shared embryological origins.

Mechanism of referral
There are several proposed mechanisms for referred pain. Currently there is no definitive consensus regarding which is correct. The cardiac general visceral sensory pain fibres follow the sympathetics back to the spinal cord and have their cell bodies located in thoracic dorsal root ganglia 1-4(5). As a general rule, in the thorax and abdomen, general visceral afferent (GVA) pain fibres follow sympathetic fibres back to the same spinal cord segments that gave rise to the preganglionic sympathetic fibres. The central nervous system (CNS) perceives pain from the heart as coming from the somatic portion of the body supplied by the thoracic spinal cord segments 1-4(5). Also, the dermatomes of this region of the body wall and upper limb have their neuronal cell bodies in the same dorsal root ganglia (T1-5) and synapse in the same second order neurons in the spinal cord segments (T1-5) as the general visceral sensory fibres from the heart. The CNS does not clearly discern whether the pain is coming from the body wall or from the viscera, but it perceives the pain as coming from somewhere on the body wall, i.e. substernal pain, left arm/hand pain, jaw pain.
### Chart of referred symptoms

<table>
<thead>
<tr>
<th><strong>Lungs</strong></th>
<th><strong>Large Intestine</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Liver, stomach, oesophagus, ribs, sternum, costal cartilage</td>
<td>Acute or chronic low back pain</td>
</tr>
<tr>
<td>Rib 1 – stellate ganglion</td>
<td>Sciatica left (venous circulation problems)</td>
</tr>
<tr>
<td>T 1-4</td>
<td>Sciatica right (caecum)</td>
</tr>
<tr>
<td>Cervical spine</td>
<td>Varicose veins – left</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Liver</strong></th>
<th><strong>Kidneys</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>C 4-5 right of bilateral</td>
<td>T 6-7</td>
</tr>
<tr>
<td>Right scapula</td>
<td>T 10-12</td>
</tr>
<tr>
<td>Right glenohumeral joint</td>
<td>T11-12 costovertebral</td>
</tr>
<tr>
<td>Cervical/brachial plexus and fascia</td>
<td>Inferior navicular (K2 acupuncture point)</td>
</tr>
<tr>
<td>T 7-10, Ribs 7-10 right, costovertebral joints</td>
<td></td>
</tr>
<tr>
<td>Cranial base restriction – right</td>
<td></td>
</tr>
<tr>
<td>Sciatica left – venous hepatic origin</td>
<td></td>
</tr>
<tr>
<td>Sciatica right – related to hepatic fascia, right kidney, ascending colon</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Stomach</strong></th>
<th><strong>Jejunoileum</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Cervical spine – left with left sternoclavicular joint</td>
<td>T10-12</td>
</tr>
<tr>
<td>Glenohumeral periarthritis – left</td>
<td>Acute of chronic low back pain</td>
</tr>
<tr>
<td>T 6-11</td>
<td>Sciatica left (venous circulation problems)</td>
</tr>
<tr>
<td>T 6 – left costovertebral (stomach dermatome)</td>
<td>Joint pains in lower limbs</td>
</tr>
<tr>
<td>Rib 7 – right</td>
<td></td>
</tr>
<tr>
<td>T12 – L3 (crura)</td>
<td></td>
</tr>
<tr>
<td>Sacroiliac – left (related to L1)</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Duodenum</strong></th>
<th><strong>Bladder</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>T12-L1 (right &gt; left)</td>
<td>L 2-3 – associated with incontinence</td>
</tr>
<tr>
<td></td>
<td>Sacrococcygeal – associated with feet</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Gallbladder</strong></th>
<th><strong>Female reproductive system</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>C 4-6 left</td>
<td>Lumbosacral – urogenital problems</td>
</tr>
<tr>
<td>C4 transverse process</td>
<td>Knee – genitocrural nerve</td>
</tr>
<tr>
<td>T 7-9 right costovertebral joint</td>
<td>C 2-4 – hormone problems, via</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Coccyx</strong></th>
<th><strong>Female reproductive system</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Refers to bladder and uterus</td>
<td>Lumbosacral – urogenital problems</td>
</tr>
<tr>
<td></td>
<td>Knee – genitocrural nerve</td>
</tr>
<tr>
<td></td>
<td>C 2-4 – hormone problems, via</td>
</tr>
<tr>
<td></td>
<td>hypothalamic-pituitary axis</td>
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</table>

### Convergent-projection

Convergent projection proposes that afferent nerve fibres from tissues converge onto the same spinal neuron. This large or chronic sensory input can precipitate an ‘overflow’ to other nerves sharing that spinal segment, causing pain in other areas of the body that share that spinal segment. This explains why referred pain is believed to be segmented in much the same way as the spinal cord. Additionally, experimental evidence shows that when local pain (pain at the site of stimulation) is intensified the referred pain is intensified as well.

Criticism of this model arises from its inability to explain why there is a delay between the onset of referred pain after local pain stimulation. Experimental evidence also shows that referred pain is often unidirectional. For example stimulated local pain in the anterior tibial muscle causes referred pain in the ventral portion of the ankle; however referred pain
moving in the opposite direction has not been shown experimentally. Lastly, the threshold for
the local pain stimulation and the referred pain stimulation are different, but according to this
model they should both be the same.

**Convergence-facilitation**

Convergence facilitation believes that the internal organs are insensitive to stimuli and that
non-nociceptive afferent inputs to the spinal cord created what he termed "an irritable focus". This
focus caused some stimuli to be perceived as referred pain. However these ideas did
not gain widespread acceptance from critics due to its dismissal of visceral pain.

Recently this idea has regained some credibility under a new term, central sensitization. Central
sensitization occurs when neurons in the spinal cord's dorsal horn or brainstem
become more responsive after repeated stimulation by peripheral neurons, so that weaker
signals can trigger them. The delay in appearance of referred pain shown in laboratory
experiments can be explained due to the time required to create the central sensitization.

**Axon-reflex**

Axon-reflex suggests that the afferent fibre is bifurcated before connecting to the dorsal horn. Bifurcated
fibres do exist in muscle, skin, and intervertebral discs. Yet these particular
neurons are rare and are not representative of the whole body. Axon-Reflex also does not
explain the time delay before the appearance of referred pain, threshold differences for
stimulating local and referred pain, and somatosensory sensibility changes in the area of
referred pain.

**Hyper-excitability**

Hyper-excitability hypothesizes that referred pain has no central mechanism. However, it
does say that there is one central characteristic that predominates. Experiments involving
noxious stimuli and recordings from the dorsal horn of animals revealed that referred pain
sensations began minutes after muscle stimulation. Pain was felt in a receptive field that was
some distance away from the original receptive field. According to hyper-excitability, new
receptive fields are created as a result of the opening of latent convergent afferent fibres in
the dorsal horn. This signal could then be perceived as referred pain.

Several characteristics are in line with this mechanism of referred pain, such as dependency
on stimulus and the time delay in the appearance of referred pain as compared to local pain. However, the appearance of new receptive fields, which is interpreted to be referred pain,
conflicts with the majority of experimental evidence from studies including studies of healthy
individuals. Furthermore, referred pain generally appears within seconds in humans as
opposed to minutes in animal models. Some scientists attribute this to a mechanism or
influence downstream in the supraspinal pathways. Neuroimaging techniques such
as PET scans or fMRI may visualize the underlying neural processing pathways responsible
in future testing.

**Thalamic-convergence**

Thalamic convergence suggests that referred pain is perceived as such due to the
summation of neural inputs in the brain, as opposed to the spinal cord, from the injured area
and the referred area. Experimental evidence on thalamic convergence is lacking. However,
pain studies performed on monkeys revealed several pathways converging on both subcortical and cortical neurons.

Referred pain can be indicative of nerve damage. A case study done on a 63-year-old man with a sustained injury during his childhood developed referred pain symptoms after his face or back was touched. After even a light touch, there was shooting pain in his arm. The study concluded that the reason for this man's pain was possibly due to a neural reorganization which sensitized regions of his face and back after the nerve damage occurred. It is mentioned that this case is very similar to what phantom limb syndrome patients suffer. This conclusion was based on experimental evidence gathered by V. Ramachandran in 1993, with the difference being that the arm that is in pain is still attached to the body.

**Orthopaedic diagnosis**

From the above examples one can see why understanding of referred pain can lead to better diagnoses of various conditions and diseases. In 1981 physiotherapist Robin McKenzie described what he termed centralization. He concluded that centralization occurs when referred pain moves from a distal to a more proximal location. Observations in support of this idea were seen when patients would bend backward and forward during an examination.

Studies have reported that the majority of patients that centralized were able to avoid spinal surgery via isolating the area of local pain. However, the patients that did not centralize had to undergo surgery to diagnose and correct problems. As a result of this study there has been a lot of research into the elimination of referred pain through certain body movements.

One example of this is referred pain in the calf. McKenzie showed that the referred pain would move closer to the spine when the patient bent backwards in full extension a few times. More importantly, the referred pain would dissipate even after the movements were stopped.

**General diagnosis**

As with myocardial ischaemia referred pain in a certain portion of the body can lead to a diagnosis of the correct local centre. Somatic mapping of referred pain and the corresponding local centres has led to various topographic maps being produced to aid in pinpointing the location of pain based on the referred areas. For example local pain stimulated in the oesophagus is capable of producing referred pain in the upper abdomen, the oblique muscles, and the throat. Local pain in the prostate can radiate referred pain to the abdomen, lower back, and calf muscles. Kidney stones can cause visceral pain in the ureter as the stone is slowly passed into the excretory system. This can cause immense referred pain in the lower abdominal wall.

Further, recent research has found that ketamine, a sedative, is capable of blocking referred pain. The study was conducted on patients suffering from fibromyalgia, a disease characterized by joint and muscle pain and fatigue. These patients were looked at specifically due to their increased sensitivity to nociceptive stimuli. Furthermore, referred pain appears in a different pattern in fibromyalgic patients than non-fibromyalgic patients. Often this difference manifests as a difference in terms of the area that the referred pain is found (distal vs. proximal) as compared to the local pain. The area is also much more exaggerated owing to the increased sensitivity.