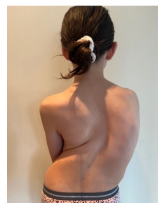




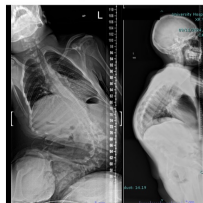
Objectives



Scoliosis Management in Children and Young People with Neurodisability



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- To understand when it is appropriate to refer to spinal surgeons
- An outline of support options offered by spinal surgery and rationale for decision making
- How often to repeat spinal x-rays in scoliotic patients
- What we should be doing as Community Paediatricians to support around scoliosis
- Prognosis of scoliosis in this patient cohort

Neurodisability



- Broad term used to describe a group of conditions that affect the central nervous system (CNS) or peripheral nervous system (PNS), leading to long-term functional impairments in movement, cognition, communication, behavior, and other aspects of daily life.
- Usually present from birth or develop in early childhood and are often permanent, though the degree of disability can vary widely.
- Examples: Cerebral Palsy, Spina Bifida, Muscular Dystrophies, Intellectual Disabilities, Autistic Spectrum Disorder, Spinal Muscular Atrophy, Acquired Brain Injuries, Epilepsy...

Neurodisability - Impact



- **Mobility:** Challenges with walking, balance, or posture.
- **Communication:** Difficulty speaking, understanding, or using alternative communication methods.
- **Learning:** Cognitive impairments or difficulties with processing information.
- **Behaviour:** Emotional regulation, social interaction, or behavioural challenges.
- **Health and Function:** Issues like epilepsy, scoliosis, feeding difficulties, or respiratory problems.

Neuromuscular



- The term neuromuscular refers to the interaction between the nervous system (central and peripheral) and the muscles, which work together to produce movement, maintain posture, and perform other motor functions.
- Neuromuscular:
 - Specifically refers to disorders affecting the nerves, muscles, or their connection.
 - Focuses on motor function and movement-related impairments.
- Neurodisability:
 - Broader term encompassing neurological conditions causing developmental and functional impairments (e.g., cognition, behaviour, and mobility).
 - May include neuromuscular disorders as a subset.

Scoliosis

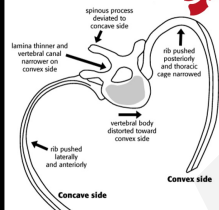
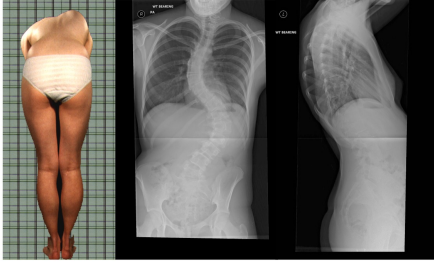


- The term **scoliosis** originates from the Greek word **σκολίωσις (skoliosis)**, which means "crookedness" or "bending." It is derived from the root **σκολιός (skolios)**, meaning "curved" or "twisted."
- In medical terms, scoliosis refers to a **lateral curvature of the spine**, often accompanied by a rotational deformity of the vertebrae.
- It is a **three-dimensional spinal deformity** that can present as an **S-shaped** or **C-shaped** curve when viewed from behind.
- Rotational and "collapsing" neuromuscular imbalance elements to the deformity

Scoliosis



Rib Hump



Scoliosis



Scoliosis



Scoliosis Classification

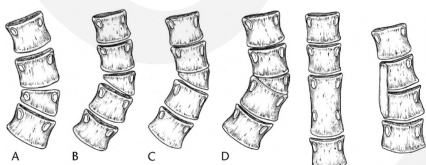


- Early onset < 10 years
 - Idiopathic, Congenital, Neuromuscular, Syndromic
- Late onset > 10 years
- Adult onset
- Main Causes:
 - Idiopathic (80-85%), Congenital, Neuromuscular, Syndromic, Degenerative, Trauma, Infection, Tumour

Scoliosis Classification



- Failure Formation / Segmentation / Both

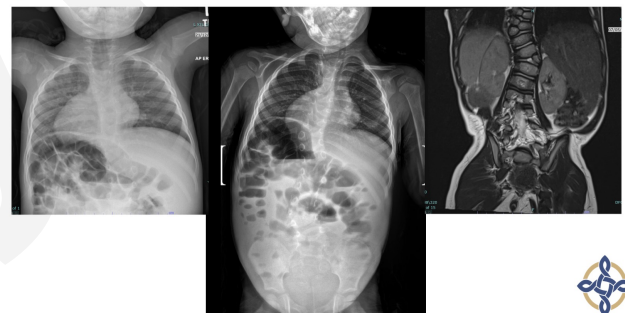


- V - Vertebral
- A - Anorectal
- C - Cardiac
- T - Tracheo
- E - Esophageal
- R - Renal
- L - Limbs

FIGURE 1. Schematic representation of formation failures. A. Wedge vertebra. B. Fully segmented hemivertebra. C. Partially segmented hemivertebra. D. Unsegmented hemivertebra. Reproduced with permission from Hedegquist and Emans.¹⁴

FIGURE 2. Schematic representations of failures of segmentation. A. Block vertebra. B. Bar. C. Bar with costalateral hemivertebra. Reproduced with permission from Hedegquist and Emans.¹⁴

Scoliosis Classification



Scoliosis Classification



Incidence of NM Scoliosis



BOX 3-1 Etiologic Classification of Scoliosis

Idiopathic	Neuromuscular—cont'd
Early-onset	Spinocerebellar degeneration
Late-onset	Cerebral palsy
Adult	Lower motor neuron
Congenital	Poliomyelitis
Osteogenic (vertebral anomaly)	Myopathy
Anomalous formation	Congenital hypotonia
Wedge vertebra	Duchenne muscular dystrophy
Hemivertebra	Developmental Syndromes
Anomalous segmentation	Skeletal dysostosis (e.g.,
Unilateral bar	neurofibromatosis)
Fused ribs	Skeletal dysplasia (e.g., osteogenesis
Fused vertebra	imperfecta)
Neuropathic (spinal dysraphism)	Tumor Associated
Tethered cord	Vertebral
Chiari malformations	Osteoblastoma
Syringomyelia	Osteoid osteoma
Durasteratomyelia	Intraspinous
Meningocele/myelomeningocele	Intramedullary (e.g.,
Neuromuscular	astrocytoma)
Neuropathic	Extramedullary (e.g.,
Upper motor neuron	neurofibroma)

BOX 11-1 Neuromuscular Disorders Associated with Scoliosis and Their Classification

Neuropathic
Upper motor neuron
Cerebral palsy
Spinocerebellar degeneration
Friedreich's ataxia
Charcot-Marie-Tooth
Roussy-Levy
Syringomyelia
Spinal cord tumor
Spinal cord trauma
Lower motor neuron
Poliomyelitis and other viral myelitis
Traumatic
Spinal muscle atrophy
Werdnig-Hoffmann
Kugelberg-Welander
Dysautonomia
Myopathic
Arthrogryposis
Muscular dystrophy
Duchenne
Limburg
Facioscapulohumeral
Fiber type disproportion
Congenital hypotonia
Myotonia dystrophica



Diagnosis

Cerebral palsy (2 limbs involved)	25%
Myelodysplasia (lower lumbar)	60%
Spinal muscle atrophy	67%
Friedreich ataxia	80%
Cerebral palsy (4 limbs involved)	80%
Duchenne muscular dystrophy	90%
Myelodysplasia (thoracic level)	100%
Traumatic paralysis (<10 years)	100%

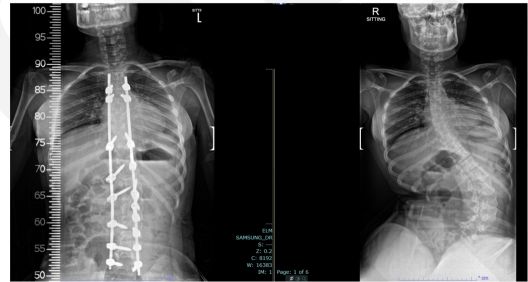


Goals of Scoliosis Mx

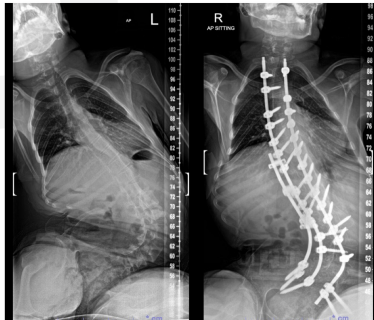
- Prevent curve progression and correct deformity
- Preserve or enhance pulmonary and cardiovascular function
- Alleviate pain and discomfort
- Balance the spine above a level pelvis
- Correct any pelvic obliquity
- Improve sitting position and reduce any pressure area issues
- Reduce any costopelvic impingement
- Optimize functional ability / reduce trunk fatigue
- Minimize psychological and social impact
- Avoid or minimize complications



Goals Scoliosis Mx



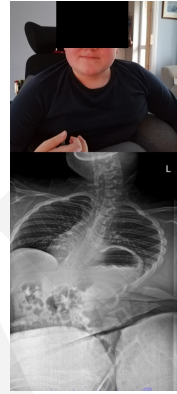
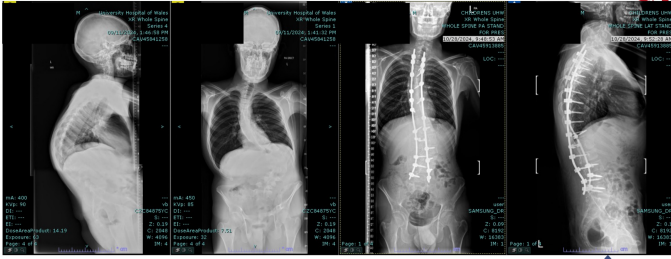
Goals Scoliosis Mx



Goals Scoliosis Mx



Goals Scoliosis Mx



Goals Scoliosis Mx



Who / When to Refer

- 1. Significant Curve Progression**
 - Rapid Curve Progression: Cobb angle $>10^\circ$ increase within 6-12 months.
 - Large Cobb Angles: Cobb angle $>40^\circ$ in children and adolescents who are still growing.
 - Severe Curves in Young Patients: Cobb angle $>20-25^\circ$ in young children (e.g., under 10 years old) warrants early referral due to a higher risk of progression.
- 2. Impact on Function and Quality of Life**
 - Seating and Posture Issues: Difficulty maintaining upright posture in a wheelchair or adaptive seating system.
 - Pain or Discomfort: Development of significant back pain or discomfort.
 - Difficulty with Hygiene and Caregiving: Challenges in personal hygiene, dressing, or caregiving due to severe spinal deformity.
- 3. Respiratory Compromise**
 - Scoliosis causing restrictive lung disease or interfering with respiratory function.
 - Frequent respiratory infections or reliance on ventilatory support may also indicate a need for evaluation.



When / Who to Refer

- 4. Surgical Feasibility and Timing**
 - Consider referral early enough to allow for surgical planning before curves become so severe that surgical correction is less feasible or higher risk (e.g., Cobb angles $>90^\circ$).
 - Scoliosis with significant spinal stiffness or secondary complications may require earlier assessment.
 - The patient must be fit for surgery / prognosis of underlying condition...**
- 5. Pelvic Obliquity and / or Costo Pelvic Impingement**
 - Scoliosis with associated pelvic obliquity causing uneven sitting surfaces, skin breakdown, or asymmetry in weight-bearing.
- 6. Neurological Decline**
 - Onset of worsening neurological symptoms, such as changes in muscle tone, spasticity, or motor function, potentially linked to the scoliosis.



When / Who to Refer

- 5. Coexisting Conditions**
 - Hip Dysplasia: Often coexists with neuromuscular scoliosis and may require combined orthopedic and spinal management.
 - Spinal Cord Issues: Conditions like tethered cord syndrome or syringomyelia should be assessed for neurosurgical input.
- 6. Prior Conservative Measures Have Failed**
 - Bracing, seating modifications, or physical therapy are not halting progression or adequately managing symptoms.
 - TIMING FOR ALL PAEDIATRIC SPINAL DEFORMITY REFERRALS IS SET TO BE AGREED BY SWSN AT <3 MONTHS...**



SWSN Guidelines - DRAFT



**SOUTH WALES SPINAL NETWORK (SWSN)
CLINICAL PATHWAY (CP)**

Paediatric Spinal Deformity

4. Primary Care & Community Care, Initial Assessment, Management and Triage

CYP with spinal deformity usually present to their GP services initially. The initial presentation may be based on the identification of a possible deformity by the patient, friend or family member. Others may present with pain, usually localised to the spine, but on occasions be associated with sciatic or nerve related symptoms e.g. pain radiating down the legs or associated sensory symptoms i.e. numbness, pins and needles.

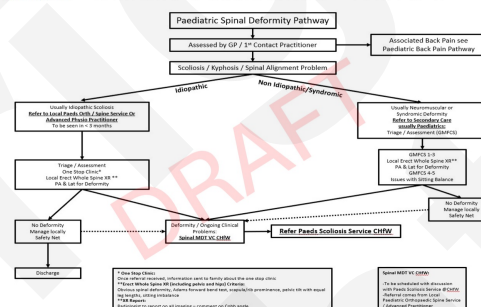
The initial presentation assessment for back pain in CYP can occur in ED or as self-referrals to physiotherapy as well.

The initial assessment should be focussed on identifying and triaging patients into streams depending on the presence or absence of red flags and symptoms/ signs suggestive of serious pathology. Refer to SWSN CP03 for more detail on the different routine and urgent referral streams. Refer to appendix 1 for detail on red flags.

This pathway concerns patients where the primary issue is suspected abnormal spinal curvature –

A. CYP with suspected abnormal spinal curvature without back pain which are non-syndromic or idiopathic should be referred to the appropriate local services i.e. APP's, paediatric orthopaedic or spine service to be assessed in 3 months. HBs should work towards AHP delivery of spinal deformity assessment with competency framework. See annex 1

B. CYP with syndromic conditions are usually under the care of the local paediatric services. If not, then they should be referred to the local paediatric services for triage and to be assessed within 3 months.



Non-Surgical Options

- **Observation and Monitoring**
 - Regular clinical and radiographic follow-ups: assess scoliosis progression, monitor growth and neurological status.
- **Bracing**
 - Custom-made orthoses to support posture and potentially slow curve progression. Not curative (in neuromuscular deformity) but may help delay surgery in younger patients. Provides trunk support for improved seating and daily activities.
- **Seating Modifications**
 - Collaboration with occupational therapists and physiotherapists to improve wheelchair or seating systems (e.g., molded seats, lateral supports). Helps maintain a functional sitting position, improving comfort and reducing risk of pressure sores and skin breakdown.
- **Pain and Symptom Management**
 - Advice on pain management strategies, incl. medication or non-invasive therapies. Improves quality of life, particularly in children with advanced scoliosis where surgery is not possible.
- **Education and Support**
 - Informing Families: educate about scoliosis, progression risks, importance regular follow-ups.



Spinal Surgery

- **Growth-Friendly Surgical Techniques**
 - Growing rods for younger children to allow continued spinal and thoracic growth.
 - Vertical Expandable Prosthetic Titanium Rib (VEPTR) for children with thoracic insufficiency syndrome.
 - Control progression of scoliosis while accommodating growth.
 - Maintains lung development and prevents severe deformity.
- **Definitive Spinal Fusion**
 - Correct and stabilize the curve: typically in adolescents or after growth is complete.
 - Provides long-term correction and prevents further progression.
 - Improves trunk balance, seating, and quality of life.
- **Pelvic Fixation**
 - Fusion extended to the pelvis for scoliosis with significant pelvic obliquity.
 - Stabilizes the pelvis, improving seating and pressure distribution.



X-Rays – Monitoring the Curve

- **Initial Assessment**
 - Baseline X-ray (standing or sitting) when scoliosis is suspected or identified, or if signs of progression (e.g., worsening posture, changes in seating tolerance).
- **During Growth Phases**
 - Rapid Growth Phases (Prepubertal or Adolescents): X-rays every 6 months to monitor for curve progression, especially if the Cobb angle is $>20^\circ$ or if the child is undergoing conservative management (e.g., bracing).
 - Slow Growth Phases (Stable Periods): X-rays every 12 months if the curve appears stable and there is no significant growth spurt.
- **Post-Surgical Monitoring**
 - Postoperative X-rays: Typically performed at 6 weeks, 3 months, 6 months, and 12 months postoperatively.
 - Long-term follow-up: Annually or biannually, depending on the stability of the surgical correction and the presence of any complications (e.g., implant loosening or pseudarthrosis).
- **Severe or Rapidly Progressive Scoliosis**
 - Curves progressing rapidly or approaching surgical thresholds, X-rays may be needed every 3-6 months, especially in young children with neuromuscular scoliosis.
- **Non-Ambulant Patients with Stable Curves**
 - Small, non-progressive curve and skeletally mature or nearing skeletal maturity, consider reducing X-rays to every 1-2 years or when clinically indicated (e.g., change in posture, pain, or functional decline).



Community Paed Role in Scoliosis

- **Early Detection and Monitoring**
 - Assess for scoliosis during regular health check-ups, particularly in children with known neurodisabilities (e.g., cerebral palsy, muscular dystrophy, spina bifida).
 - Look for clinical signs such as: Asymmetrical shoulders or hips. Rib prominence. Difficulty with seating posture. Changes in functional ability (e.g., walking or transferring).
 - Growth Monitoring: Measure and track height, weight, and growth patterns to identify periods of rapid growth when scoliosis progression is more likely.
 - Referral for Imaging: Arrange for spinal X-rays if scoliosis is suspected or progression is noted.
- **Early Referral to Specialists**
 - Significant or progressive spinal curves (as detailed above).
 - Rapid growth or signs of worsening deformity.
 - Impact on function, posture, or respiratory health.
 - Persistent pain or discomfort related to scoliosis.



Community Paed Role in Scoliosis



- **Coordination of Multidisciplinary Care**
 - Work closely with spinal surgeons, physiotherapists, occupational therapists, speech and language therapists, and neurologists to ensure comprehensive care.
 - Seating and Equipment Needs: Coordinate with therapists to ensure appropriate adaptive seating, wheelchairs, and assistive devices to support posture and function.
 - Nutrition Support: Involve dietitians to optimize nutrition, particularly if scoliosis or neurodisability affects feeding or growth.
- **Symptom Management**
 - Pain Management: Identify and manage pain with appropriate pharmacological or non-pharmacological strategies.
 - Tone and Spasticity Management: Collaborate with neurologists, paediatric ortho or rehabilitation specialists for managing spasticity (e.g. baclofen, Botox injections).
 - Respiratory Care: Monitor for signs of respiratory compromise due to scoliosis and refer to respiratory specialists when needed.

Community Paed Role in Scoliosis



- **Education and Support for Families**
 - Navigating Healthcare Services: Guide families through the referral process and assist with accessing specialized care.
 - Psychosocial Support: Provide emotional and psychological support to the child and their caregivers, acknowledging the challenges of living with scoliosis and neurodisability.
 - School and Community Support: Advocate for necessary adjustments at school or in the community to accommodate the child's physical and educational needs.
- **Long-Term Follow-Up**
 - Regular Monitoring: Continue to monitor growth, posture, and function in collaboration with specialists.
 - Addressing Coexisting Conditions: Manage other aspects of the child's neurodisability (e.g., epilepsy, orthopedic issues, nutritional concerns).
 - Transition to Adult Services: Prepare the family for transitioning the child to adult care services, ensuring continuity of management.

Prognosis - Factors



- **Underlying Neurological Condition**
 - Children with severe neuromuscular disorders (e.g., Duchenne muscular dystrophy, spinal muscular atrophy, severe cerebral palsy) often have more aggressive scoliosis progression.
 - Conditions with spasticity (e.g., cerebral palsy) or hypotonia (e.g., muscular dystrophies) predispose to more significant deformities.
- **Skeletal Maturity**
 - Younger children are at greater risk of progression as still growing.
- **Curve Severity at Diagnosis**
 - Larger curves (>40-50° Cobb angle) progress more quickly and are more likely to cause complications.
- **Functional Status**
 - Ambulatory children often have a slower progression and better functional outcomes compared to non-ambulatory children.
- **Timeliness of Intervention**
 - Early detection and management, including bracing or surgery, can prevent severe deformity and improve outcomes.

Summary – Objectives!



- To understand when it is appropriate to refer to spinal surgeons
- An outline of support options offered by spinal surgery and rationale for decision making
- How often to repeat spinal x-rays in scoliotic patients
- What we should be doing as Community Paediatricians to support around scoliosis
- Prognosis of scoliosis in this patient cohort

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