GUIDELINE

Congenital Hemivertebrae

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Aim

To identify and manage congenital hemivertebra in a newborn.

Risk

Risk to patient: Non or delayed identification may increase the risk of missing other associated problems and progressive worsening of spine deformity without the opportunity for surveillance and early intervention.

Risk to institution: Suboptimal clinical care and possible medico-legal risk attributed to the same.

Background

Hemivertebrae (HV) is a condition where one half of the vertebra completely fails to form. It is the most common aetiology of congenital scoliosis, lordosis or kyphosis (1,2). HV may be fully segmented, partially segmented, or unsegmented and are more common in females. Exact aetiology is unclear and involves both genetic and environmental factors (3).

The type and location of HV can affect the likelihood and the rate of progression of spine deformity (4). The degree of spine deformity tends to worsen as the child grows (5). The rate at which scoliosis or spine deformity develops depends on many factors including the type of HV, number and position of HV in the spine, ipsilateral v/s bilateral in case of multiple HV and age of the patient (4).

Antenatal 3D ultrasound is potentially a useful modality to detect the exact location, type, number of affected vertebra and associated anomalies and hence determine

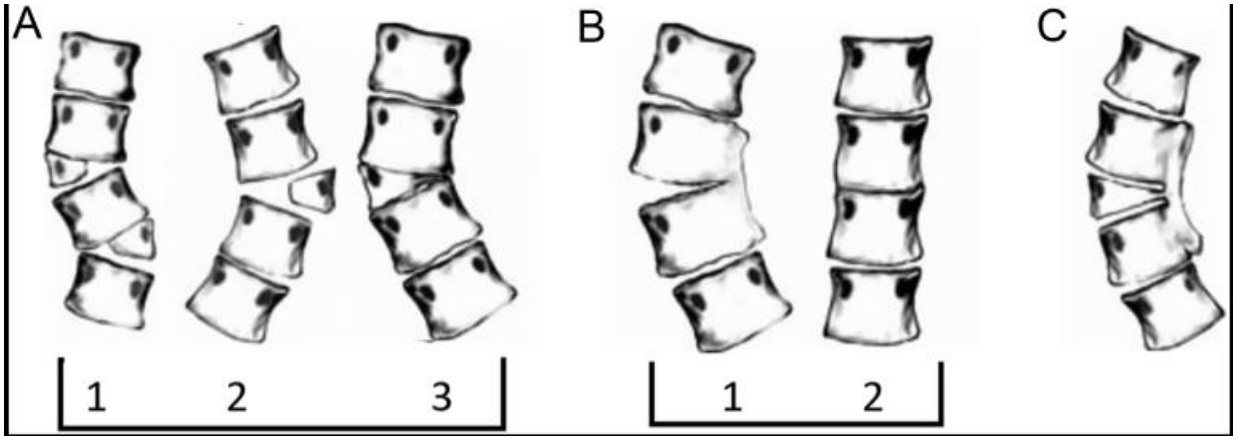
prognosis in hemivertebrae (6). Isolated foetal HV carry a good prognosis and associated anomalies and presence of oligohydramnios reduces survival significantly (7). Diagnosis of foetal HV should be confirmed by foetal MRI, especially to evaluate associated CNS abnormalities and assist with appropriate neonatal management and parental counselling (8).

In some cases, scoliosis can progress rapidly and hence can benefit from early orthopaedic referral. Surgical intervention is most commonly used when the HV is located from the thoracolumbar to lumbosacral junction (9).

Even though most HV occur in isolation, a significant number could be associated with GIT, CNS, renal, other vertebral and cardiovascular anomalies. Segmentation anomalies may be associated with syrinx, diastematomyelia and tethered cord (10). They can also be part of various genetic syndromes: Jarcho-Levin syndrome (AR, fused vertebrae, scoliosis, abnormal rib alignment), Klippel-Fiel syndrome (AR/ AD, fused cervical vertebrae), VACTERL (sporadic) and OEIS complex (sporadic; omphalocele, cloacal exstrophy, imperforate anus and spinal defects) (11).

The mainstay of treatment remains early diagnosis and intervention before severe curvature and deformity occur. Hence, early and regular review by the orthopaedic team is essential.

Variations of Hemivertebrae



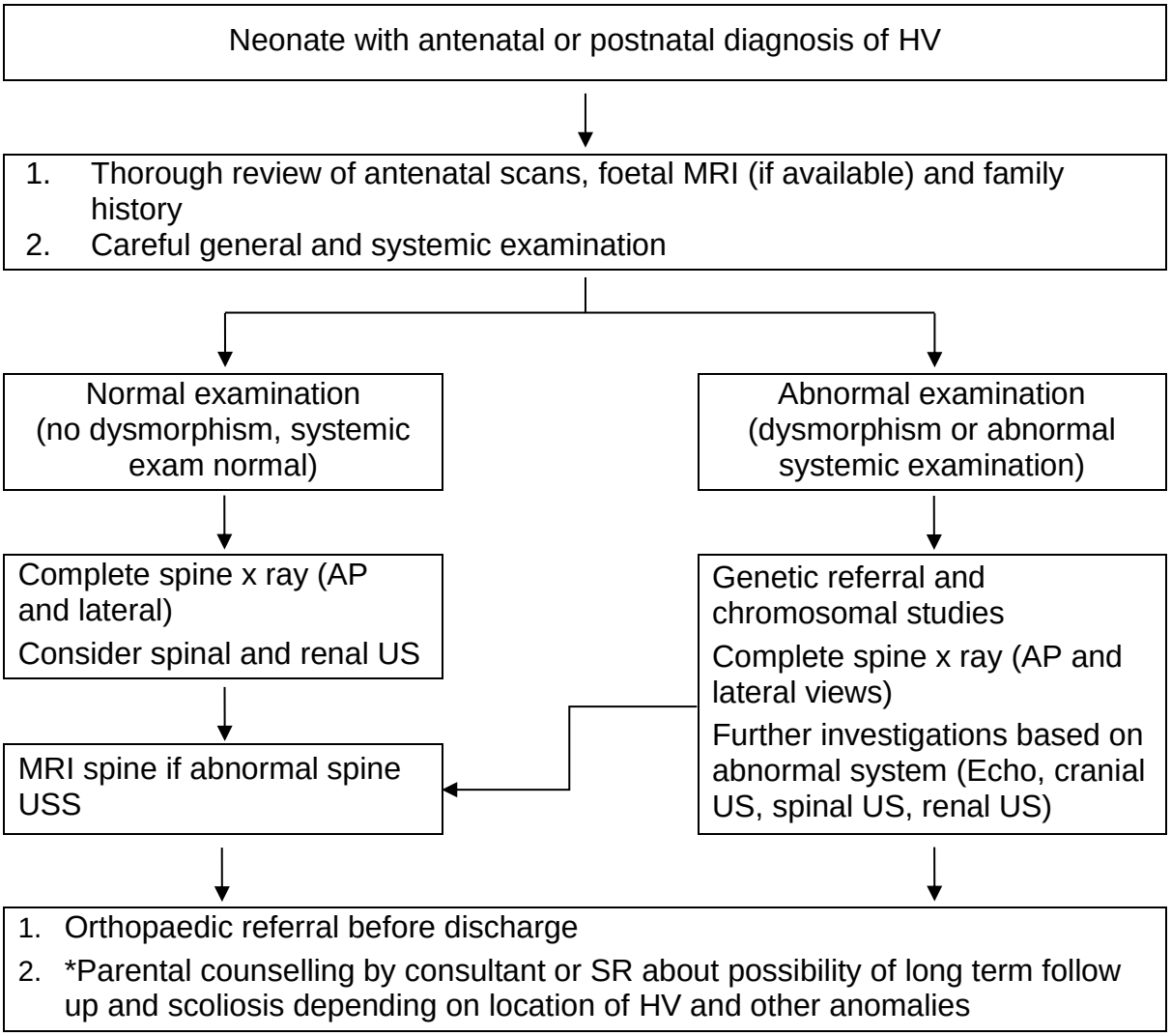
(A): failure of formation (A1: semi-segmented, A2: fully segmented; A3: wedge vertebra), (B) failure of segmentation (B1: bar, B2: block), (C) mixed deformities (*12*)

Natural history of congenital scoliosis (to assist counselling)

1. Age: Progression is rapid before age 5 years and adolescence (11-14 years). Curves clinically present before 10yrs have poor prognosis, especially if present in the first year of life.
2. Location of the apex: Upper thoracic have slowest progression> midthoracic> thoracolumbar (fastest).

3. Type of anomaly: Worst prognosis: unilateral bar with contralateral HV and most benign: complete block/ incarcerated HV. Higher progression: presence of >1 HV, thoracolumbar hemi-metameric shifts, presence of bar or fused rib (simulates a tether)
4. Curve characteristics: two unilateral curves: deep malformation vs contralateral curves: balance. Progression unlikely if Cobb's angle<25º

Management of a Neonate with Antenatal / Postnatal Diagnosis of HV



(Jape G, Rao S, Minutillo C, Thonell S, 2011, Sebaaly 2022)

References and related external legislation, policies, and guidelines

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