CONGENITAL KYPHOSIS, EARLY MANAGMENT

Essay

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TABLE OF CONTENT

1. INTRODUCTION…….……..….1

2. EMBRYOLOGY AND GENETICS OF THE SPINE…….....3

3. ANATOMY OF THE SPINE….....19

4. BIOMECHANICS OF THE SPINE…….....33

5. CLASSIFICATION OF CONGENITAL KYPHOSIS……...38

6. CLINICAL EXAMINATION AND DIAGNOSIS……53

7. SURGICAL INTERVENTION, TIMING, INDICATION AND CONTRAINDICATION…….....78

8. SURGICAL TECHNIQUE……87

9. SUMMERY…….....128

10. REFERENCES……...130

الملخص العربى

LIST OF FIGURES

Figure 1: Normal human gastrulation. 4
Figure 2: Formation of somites. 5
Figure 3: Development of sclerotomes into the vertebral column. 7
Figure 4: Axial images of a vertebra demonstrating the stages of development of the vertebral column. [43] 10
Figure 5: Proposed embryogenesis of split cord malformations. 13
Figure 6: hemimetameric somitic shift producing a hemivertebra. 14
Figure 7: diagrammatic schemes of the different vertebral malformations. 18
Figure 8 : The vertebral column. 19
Figure 9 : IVDs and ligaments of the spine. 20
Figure 10 : post natal development of the spine curvature and the vertebrae. 23
Figure 11 : Postnatal development of the vertebral body. 25
Figure 12 : Secondary ossification centers. 26
Figure 13 : CT axial views of the cervical vertebrae of C3-7. 27
Figure 14 : IVDs. 28
Figure 15 : The position of the spinal cord and meninges in relation to the vertebral column. 30
Figure 16 : the centre of body mass. 37
Figure 17: Winter's classification. 38
Figure 18: McMaster and Singh classification. 39
Figure 19: Anterior failure of vertebral body formation. 40
Figure 20: Anterior failure of vertebral body segmentation. 41
Figure 21: mixed anomalies 42
Figure 22: The advent of 3D CT over CPSI (2D) 43
Figure 23 : Hemivertebra with posterior fusion. 44
Figure 24: Fully-segmented hemivertebra with hemilamina. 44
Figure 25: Fully-segmented hemivertebra and fully segmented bilamina. 45
Figure 26 : Fully-segmented hemivertebra and spina bifida. 45
Figure 27 : Typical variation of type of formation failure in anterior and posterior components based on 3DCT. 46
Figure 28 : Anteroposterior (AP) discordant anomaly. 47
Figure 29 : Segmentation failure only (unison). 48
Figure 30 : AP discordant segmentation anomaly 49

Figure31 : Segmentation Abnormalities in Formation Failure (butterfly). 50
Figure32 : Segmentation Abnormalities in Formation Failure (hemivertebra). 50
Figure 33: Discordant anomaly in formation failure..... 51
Figure 34: Drawings of the 2 types of complex type. 52
Figure 35: Examination in standing position 58
Figure 36: Neurological evaluation..... 61
Figure 37: (A) L4 and (B) L5 root examination. 63
Figure 38: S1 root examination..... 64
Figure 39: diagrammatic approach to a patient with congenital vertebral malformation. 65
Figure 40: Longitudinal ultrasonographic scans of the spine of a second trimester fetus with hemivertebra. 66
Figure 41: Patient positioning for ultrasound. 67
Figure 42 : Ultrasound image of a normal spinal cord (large arrows) in a newborn. 68
Figure 43 : Risser sign (the stage of skeletal maturation). 69
Figure 44: Cobb method. 70
Figure 45: Evaluation of sagittal spinal curvature in 2D images. 72
Figure 46: MRI of the thoracolumbar spine..... 74
Figure 47 : Sequence of NCS closure. 75
Figure 48: Axial MRI T2-weight images demonstrating the grading scale of the NCS. 76
Figure 49 : Prophylactic Posterior Spine Arthrodesis..... 80
Figure 50 : Prophylactic Posterior Spine Arthrodesis. 80
Figure 51 : instrumented posterior fusion. 81
Figure 52 : combined anterior and posterior fusion. 82
Figure 53: Anterior release. 90
Figure 54: Anterior Cord Decompression and Fusion. 92
Figure 55: Anterior Osteotomy and Fusion. 94
Figure 56: Anterior strut grafting. 95
Figure 57: tibial strut graft. 96
Figure 58: Anterior vascularized rib graft..... 98
Figure 59: Vascularized fibular graft. 99
Figure 60: Zielke instrumentation. 101
Figure 61: Moe facet fusion. 103
Figure 62: Posterior segmental wiring. 104
Figure 63: Different types of hooks. 105
Figure 64: pediatric pedicle screws. 106

Figure 65: Different types of pedicles..... 106
Figure 66: pedicle screws..... 107
Figure 67: posterior spinal tether effect. 110
Figure 68: Algorithm of osteotomy type based on the character of the sagittal deformity..... 112
Figure 69: marking the entry point of pedicle screws with cannulas..... 113
Figure 70: HV excision. 114
Figure 71: posterior HV resection..... 115
Figure 72: (a–e) 11-year-old boy with congenital kyphosis with tethered cord and gait disturbances. 116
Figure 73: Three Rod Technique for HV resection. 117
Figure 74: video assisted thoracoscopic surgery..... 118
Figure 75: HV excision through anterior/posterior resection. 125

INTRODUCTION

The word kyphosis was derived from the Greek word kyphos which means a hump, (-osis) a suffix denoting a process especially a disease. Spinal deformity has been recognized since ancient times, Skeletal remains and archeological artifacts of prehistoric man exhibit graphic depictions of spinal anomalies and pathological curvature. It was ***Hippocrates***, in the 5th century BC, who described it for the 1st time with scientific prudence. (1)

Congenital kyphosis is a sagittal plane deformity characterized by abnormal posterior convex curve of a segment of spine. Congenital kyphosis or kyphoscoliosis results from developmental vertebral anomalies that impair longitudinal growth anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane. (2)

The term “congenital” is slightly misleading because it implies that the curvature is apparent at birth, but this is not necessarily so. It is the vertebral anomalies that are present at birth and the clinical deformity only develops with spinal growth and may not become apparent until later childhood when the diagnosis is made radiographically. (3)

Congenital kyphosis is a rare spinal deformity that usually is progressive without surgical intervention. Progression of the deformity may lead to paraplegia and cardiopulmonary dysfunction, Therefore, prompt recognition and early treatment of this disorder is preferred. (4)

Spinal cord compression is the worst complication of a congenital kyphosis or kyphoscoliosis but only occurs in patients with an anterior failure of vertebral body formation, the greatest risk of spinal cord compression occurs when the curve apex is in the mid and upper thoracic regions. (5)

Intraspinal anomalies are associated with 37% the congenital spinal deformity cases, according to MRI. Such anomalies are significantly more common in the patients with congenital kyphosis than in the patients with congenital scoliosis [6], the most common intraspinal anomalies diagnosed with MRI are tethered spinal cord and diastematomyelia. [7]

Nonoperative treatment including full time bracing has been ineffective in controlling progressive deformity, operative correction of congenital kyphosis is still a challenging problem in the growing spine. [8]

The skill in managing a patient with a congenital kyphosis lies not just in the ability to perform major complex surgery at a late stage but primarily in recognizing those curves with a severe spinal growth imbalance at an early age and applying prophylactic surgical treatment to prevent curve progression. [3]

EMBRYOLOGY AND GENETICS

Embryology of the spine

The development of the spine includes six separate but overlapping phases. [9]

1st Phase: Gastrulation and formation of somites and notochord

Within the first 2 weeks after fertilization, the embryo undergoes several cell divisions to form a blastocystda two layered embryo suspended between the amnionic and yolk sacs, Cells on the dorsal surface adjacent to the amnionic cavity comprise the epiblast, whereas cells on the ventral surface adjacent to the yolk sac comprise the hypoblast. At this point, the embryo exhibits a craniocaudal orientation, with the prochordal plate visible as a cranial thickening. [9; 10; 11]

During the second week, the embryo undergoes gastrulation, a midline primitive streak develops at the caudal end of the embryo and elongates cranially over 3 days, occupying the midline in the caudal half of the embryo. The primitive streak subsequently becomes progressively shorter (regresses) and occupies a more caudal position in the embryo. Throughout gastrulation, coordinated cell movements convert the embryo from two layers (epiblast and hypoblast) to three layers (ectoderm, mesoderm, and endoderm). Epiblast cells migrate toward the primitive streak and invaginate through the primitive groove within the primitive streak (figure 1A). While the primitive streak is still elongating, these invaginating cells form the endoderm. Later, as the primitive streak regresses toward the caudal pole, the invaginating cells migrate between the epiblast and the newly formed endoderm to form the somitic mesoderm. The remaining epiblast cells spread out to replace the cells that have invaginated through the primitive groove and form neuroectoderm and cutaneous ectoderm. At the cranial end

of the primitive streak is Hensen's node, within which is a cranial extension of the primitive groove called the primitive pit (figure 1B). As the primitive streak regresses, cells within Hensen's node invaginate through the primitive pit to form the midline notochord. The notochord continues to elongate as the primitive streak regresses caudally and is flanked bilaterally by the newly developed somitic mesoderm; together, the notochord and somites form the axial skeleton. Both are laid down in a rostral-to-caudal direction. The caudal- most vertebrae are formed last from somitic cells derived from the caudal cell mass (the remnants of the primitive streak) and from caudal

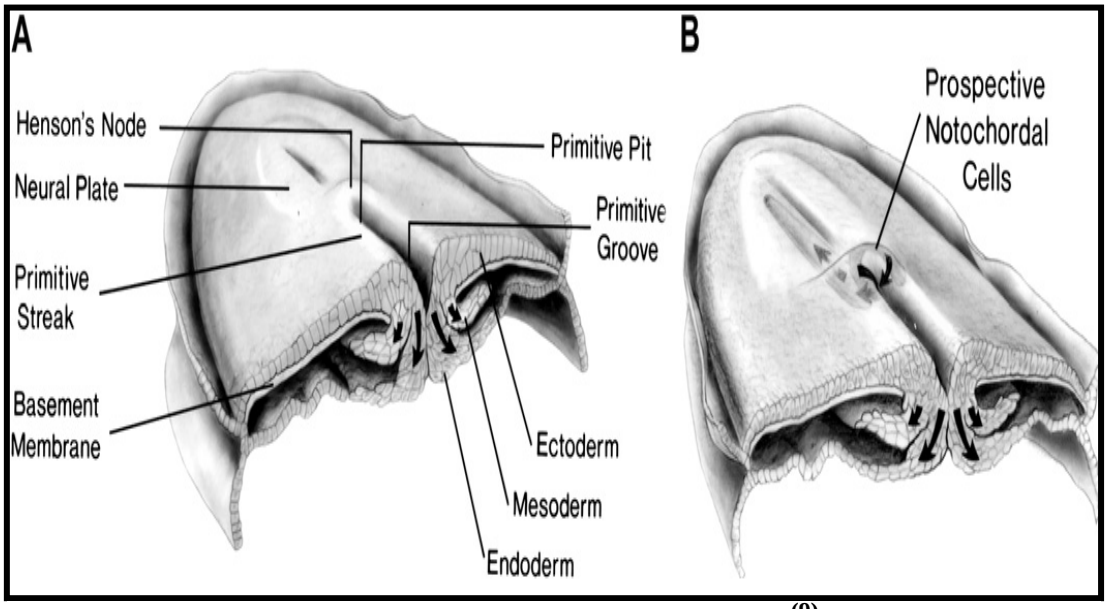

Figure 1: Normal human gastrulation. [9]

. (A) Prospective endodermal and mesodermal cells of the epiblast migrate toward the primitive streak and ingress (arrows) through the primitive groove to become the definitive endoderm and mesoderm. (B) Prospective notochordal cells in the cranial margin of Hensen's node ingress through the primitive pit during primitive streak regression to become the notochordal process.

notochordal cells derived from the posterior notochordal center immediately cranial to the caudal cell mass. [9; 10; 11; 12; 13]

2nd phase: Condensation of the somitic mesoderm to form the somites

The newly formed somitic mesoderm aggregates into discrete blocks of tissue, the somites (figure 2). The first somites form early during the third week to form the cervical vertebrae. (10; 14)

Approximately five somites are present at the time the neural tube begins to close; succeeding somites thereafter form at the level of the closing neural tube in a rostral-to-caudal sequence. (9)

The patterning of the somites is determined by the interaction of various homeobox genes and their gene products. The specification of a vertebra along the craniocaudal axis is thought to be attributable to its Hox profile the expression of various homeobox genes. Misexpression of one or

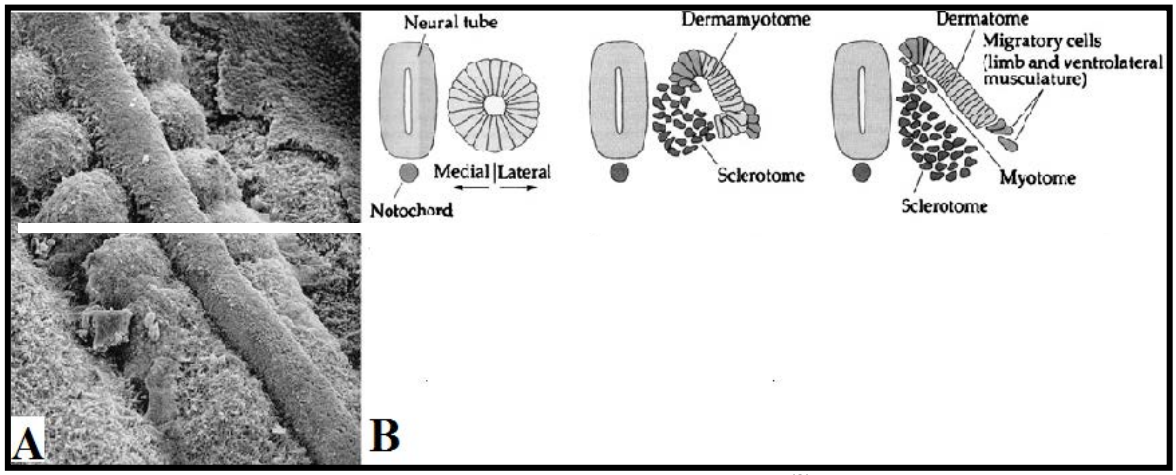

Figure 2: Formation of somites. (9)

(A) Scanning electron micrograph of a chick embryo shows the somitic mesoderm forming as blocks of tissue from the unsegmented somitic mesoderm lying on either side of the midline neural tube. (B) Somite initially forms immediately lateral to the neural tube and notochord. Reorganization within the somite forms the dorsolateral dermomyotome that gives rise to the skin and muscle, and the ventromedial sclerotome that gives rise to the vertebrae.

another homeobox gene in mice can result in cranial or caudal transformation of various vertebrae (15). The misexpression of homeobox genes in human beings could similarly account for such malformations as the occipitalized atlas, cervical ribs, and lumbarized or sacralized lumbosacral vertebrae (9; 11; 16).

The pathways involved are highly complex but involve differential expression and regulation of among others, the HOX genes and involve a variety of pathways, including Wnt, retinoic acid (RA) and fibroblast growth factor (FGF) signaling. [17]

The maximum number of somites in the human embryo is generally given as 42 to 44, although no more than 38 or 39 are required for the formation of the axial skeleton .Most of the ‘‘overage’’ is attributable to coccygeal somitic segments that disappear during subsequent growth, although a rearrangement or loss of the most cranial segments can occur as well [16]. The number and size of the somites seem to be species specific [18]. If somites are experimentally removed, the embryo is capable of compensating (regulating) to generate a normal number of somites of normal size [9; 19]

3rd phase: Formation of the sclerotome and dermomyotome

The developing somite becomes reorganized dorsoventrally into two parts, a ventral sclerotome forming the axial skeleton and a more dorsal dermomyotome forming the subcutaneous tissues and dorsal trunk musculature (figure 2B). [16; 20; 21] The sclerotome and dermomyotome are distinguished by the expression of molecular markers, with the sclerotome expressing Pax-1 and Pax-9 and the dermomyotome expressing Pax-3, Pax-7, and Myo-D [22].

The formation of the sclerotome and dermomyotome is regulated by the notochord or neural tube floor plate (the ventral portion of the neural tube); the sclerotome can be obliterated by experimentally removing the notochord or duplicated by implanting an additional dorsal notochord [15; 20].

The later development of the somites can be divided into three phases: the membranous phase, chondrification phase and ossification phase. [9; 10]

4th phase: Formation of the membranous somite and resegmentation

The membranous phase begins during the fifth embryonic week, with sclerotomal cells from each somitic pair migrating ventrally to surround the notochord and dorsally to surround the neural tube. Ventral somitic cells form the vertebral centra. Each centrum develops a craniocaudal polarity during somitogenesis, with cranial and caudal portions having a unique

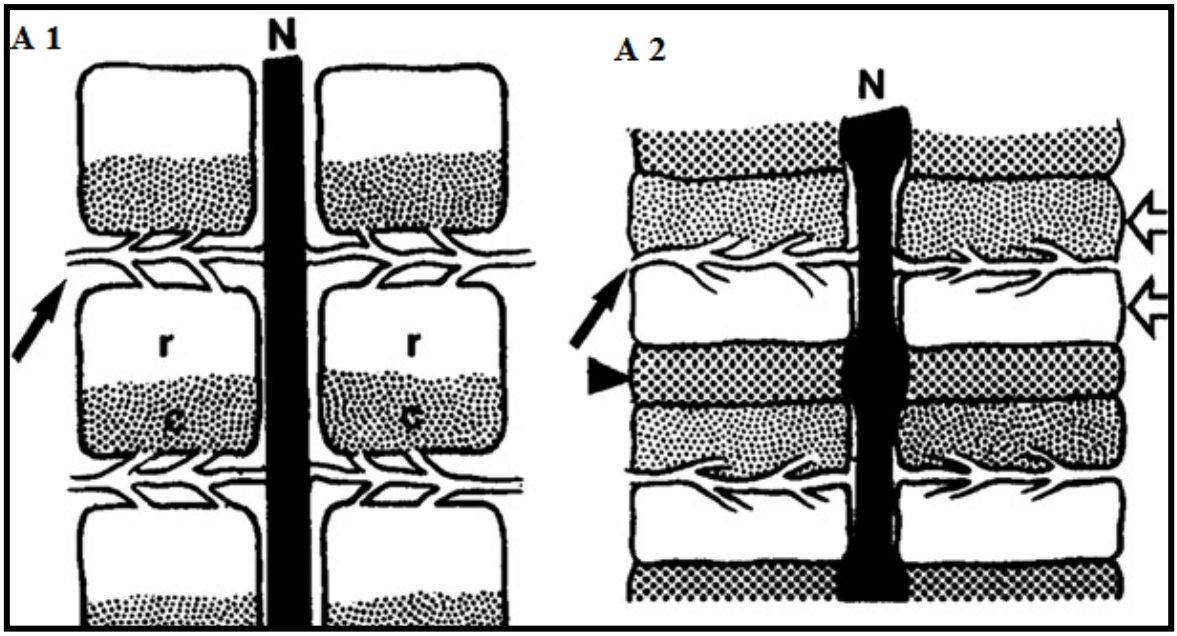

Figure 3: Development of sclerotomes into the vertebral column. [43]

Division of sclerotomes into rostral (r) and caudal (c) halves (A1) and formation of vertebral bodies (open arrows) from the fusion of the caudal half of one sclerotome and the rostral half of the adjacent sclerotome while the IVD (arrowhead) forms from the caudal half of the somite (A2).

histology and expressing a unique set of molecular markers (figure 3) [20; 22].

The cranial portion of each sclerotome is more loosely organized, whereas the caudal portion contains more densely packed cells [11]. Between the cranial and caudal portions lies a hypocellular cleft, the fissure of von Ebner. The craniocaudal organization of the sclerotome is critical to axonal outgrowth, because the outgrowth of spinal nerves at each level of the neuraxis is restricted to the more loosely organized cranial portion [22]. The

dorsal vertebral arch seems to be exclusively derived from the caudal more densely packed half of the sclerotome. [11; 21; 23]

There has been ongoing debate about whether each centrum is derived from a single sclerotome or from the fusion of the caudal and cranial halves of two adjacent sclerotomes with the hypocellular fissure of von Ebner contributing to the intervening intervertebral disc a process called resegmentation. Resegmentation was originally proposed by ***Remak*** [24] in 1855 to account for the anatomic arrangement of the vertebral centra, dorsal vertebral arch, and spinal nerves. Because the spinal nerve passes through the cranial half-sclerotome at each sclerotomal level and the posterior vertebral arch is derived from the caudal half-sclerotome, one would predict that the spinal nerve would exit cranial to the corresponding pedicle. The observation that each spinal nerve passes caudal to the corresponding pedicle could only be accounted for by resegmentation, such that the cranial loose-celled region of one sclerotome join with the caudal dense-celled region of the next more cranial sclerotome to form a single vertebral unit. [9; 10]

5th phase: Chondrification phase

Chondrification centers appear within the sclerotomes during the sixth embryonic week. Three paired chondrification centers appear for each vertebra: one pair within the vertebral centrum, a second pair dorsolaterally within the posterior vertebral arches and spinous process, and a third pair between the first two and within the transverse process and costal arch. Chondrification begins in the cervicothoracic region and extends cranially and caudally thereafter; chondrification of the vertebral centra occurs before the dorsal arches. During the chondrification phase, physaliphorous cells (large vacuolated cells containing large amount of intracytoplasmic mucoid material) of the notochord form the more centrally located nucleus pulposus and are surrounded by perinotochordal cells from the somites, which form the disc annulus (figure 4B) [23; 25].

The anterior and posterior longitudinal ligaments are formed during the chondrification phase from mesenchymal cells surrounding the cartilaginous vertebrae [9; 10; 11].

6th phase: Ossification phase

Vertebral ossification begins during the 8th embryonic week [26]and continues after birth (figure 4C). There are three primary ossification centers one for the vertebral centrum and one for each side of the dorsal vertebral arch. Within each side of the dorsal arch, the ossification centers form three independent ossification zones one each for the pedicles, lamina, and transverse processes [9]. Other authors have suggested two independent ossification centers within each vertebral centrum one dorsal and one ventral that fuse by the twentieth to twenty-fourth embryonic week. Others have suggested that as many as six primary ossification centers may be present, two forming the vertebral centrum; two forming the pedicles, lateral masses, and transverse processes; and two forming the lamina and spinous process [25].

The centra first begin to ossify at the thoracolumbar junction (T10-L1) and spread quickly to T2 to L4 vertebrae. Thereafter, ossification proceeds in a bidirectional fashion to involve more cranial and caudal vertebrae. In contrast, ossification of the dorsal arches begins simultaneously from C1 to L1 and proceeds craniocaudally thereafter. [9; 10; 11]

The centrum ossifies slightly before the dorsal arch; all ossification centers are visible by 14th weeks of gestation [26].

The expanding dorsal and ventral ossification centers meet to form the neurocentral joint of Luschka. It is important to recognize that the neurocentral joint lies not at the junction of the vertebra and pedicle but within the vertebral body. The vertebral body is therefore derived from the centrum and dorsal ossification centers, and the terms centrum and vertebral body are therefore not strictly synonymous. [27]