

Congenital and Acquired Anomalies of **the Cervical Spine**

(The Effects on Upper Cervical Stability)

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-clinically is a positive Sharp-Purser Test (validated for Ankylosing Spondylitis and R.A.)

“Knowing that a pathology exists is not enough – we need to understand the pathomechanics in order to be safe with our cervical treatments”

“Symmetry of body is a fairy tale (look at Shannon Dougherty) despite the illustrations we have seen in textbooks”

“Biomechanical models allow us to “visualize” what the joint does - however our “feel is the main determinant”

Anomalies of the C/Sp:

Developmental (ie. Congenital)

- genetic transmission;
- environmental factors (drugs, metabolism, infection & radiation exposure);
- vascular insufficiency & interuterine trauma

Acquired (trauma, pathological process)

- post-partum

- studies indicate that most congenital anomalies occurs in the earliest stages of vertebral body formation
- occurs during re-segmentation – first few weeks after conception
- associated with abnormal inter-segmental artery distribution

- during early interuterine developmental period abnormal development affects corresponding regions (especially of the same embryological block)

Summary

“If you see an anomaly in one region be on the lookout for anomalies elsewhere.”

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Summary of Vertebral Column Development

2nd Gestational Week

- mesoderm cells condense midline forming notochordal process

3rd Gestational Week

- notochordal process invaginates into notochord
- dorsal ectoderm thickens to form neural groove
- neural grooves folds, fuses, and becomes neural tube

Between 3rd & 5th Gestational Week

- segmentation (mesodermal cells condense beside notochord and segment into somites)
- medial portion of somite = sclerotome (precursor to vertebral body, ribs & ligaments)
- inter-segmental arteries (which will become the vertebral artery) exist between each sclerotome
- re-segmentation occurs (caudal ½ of one sclerotome & cranial ½ of next one fuse) forming the early vertebra
- the sclerotome migrates dorsally (posterior vertebral arch)
the sclerotome migrates anteriorly (vertebral body)
the sclerotome migrates laterally (transverse process & ribs)

6th Gestational Week Forward

- chondrification of the vertebral column begins
- primary ossification centers begin to ossify

2nd Decade of Life

- secondary ossification centers fuse

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BASILAR IMPRESSION

- ◆ upward invagination of part of bony rim of the Foramen Magnum
- ◆ floor of occiput indented by upper c/sp.
- ◆ dens may encroach on the brainstem

Primary - Congenital in nature often associated with other defects (i.e. Klippel-Feil)

Secondary - Pathological process

- Softening of the basal osseous structures
- Rickets, OA, RA, osteoporosis, osteogenesis imperfecta, neurofibromatosis

Listing of Symptoms related to Basilar Impression			
Anterior compression/ Pyramidal tract irritation	Impingement posterior by the posterior arch of the atlas	Cerebellar herniation	Vertebral artery
Muscle weakness and wasting, ataxia, spasticity, hyper-reflexia, and pathological reflexes are commonly found	Symptoms related to the dorsal columns are altered deep pressure sensibility, vibration, and proprioception	Nystagmus, ataxia, inco-ordination	Vertigo, Seizures, Syncope, mental disorientation

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ATLANTO OCCIPITAL FUSION (OCIPITALIZATION OF C1)

- ◆ Partial or complete congenital fusion of atlas and occiput
- ◆ Most common anomaly of the CV region
- ◆ Causes excessive mobility of the AA joint
- ◆ Seen: low hairlines, torticollis, short neck, restricted ROM
- ◆ 70% also have C2C3 fusion

CONGENITAL ANOMOLIES OF THE ODONTOID

- ◆ not very common in occurrence
- ◆ usually detected radiographically 2° to pain or post trauma
- ◆ exists as 4 variants (three main ones – see below)

1. Aplastic Dens:

- complete absence of the dens
- extremely rare
- concerned with instability

2. Hypoplastic Dens

- short stubby dens
- may or may not project above atlas facet articulation
- may cause AA instability

3. Os Odontoideum

- most common of the dens anomalies
- radioluscent oval ossicle with delineated and smooth cortical bone
- physically separate from the axis
- lacks additional regional malformations
- prior trauma usually exists
- associated with Klippel-Feil and Morquio's

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KLIPPEL-FEIL SYNDROME

- ◆ embryological disorder in 3rd to 5th gestational week
- ◆ women>men incidence is 1:42 000 births
- ◆ due to failure of re-segmentation of the sclerotomes
- ◆ causes block vertebrae (fusion of 2 or more levels) thus immature and smaller
- ◆ often involves occipitalization of the OA joint causing AA instability
- ◆ due to lack of re-segmentation, no IV disc exists at these levels
- ◆ if fusion C3 or above, get **Sprengel's deformity**
- ◆ Sprengel's deformity = underdeveloped, elevated scapula
- ◆ associated with organ anomalies (heart, deafness)
- ◆ appearance of a short neck, elevated scapula, low hairline

Type I - extensive cervical and upper thoracic fusion
- marked deformity of neck and back

Type II - most common (50% more prevalent)
- individuals have 1-2 paired segments fused
- often associated with hemivertebrae and OA fusion

Type III - involves the C/sp. but also the lower thoracic and lumbar

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MORQUIO'S SYNDROME

- ◆ inability to metabolize Keratin Sulfate which accumulated in brain, viscera and joints
- ◆ **short-trunk** dwarfism with large hands, coarse facial features, large L/sp lordosis, Kyphosis or Scoliosis
- ◆ may have corneal clouding but no sign of brain damage
- ◆ Hypoplasia of the Dens and ligament laxity of the transverse ligament
- ◆ Many have myelopathies by the age of 5 years

ACHONDROPLASIA

- ◆ Another form of Dwarfism
- ◆ Characteristic short-limb dwarfism due to disruption in the growth plate with abnormal endochondral bone formation causing disproportions and deformities
- ◆ No tendency toward CV instability but at risk of stenosis of the L/Sp and foramen magnum

DOWN'S SYNDROME (TRISOMY 21)

- ◆ Chromosomal disorder 1:700 live births
- ◆ Characteristic facial features, congenital heart disease, mental retardation, and ligamentous laxity
- ◆ Instability of OA joint (61%) and of the AA joint (20%)
- ◆ Dens anomalies not uncommon with Down's

PIERRE ROBIN SYNDROME

- ◆ Occurs due to proper development of the 1st branchial apparatus (face and neck region)
- ◆ Anomalies in the facial region have high correlation with cervical anomalies
- ◆ Micrognathia (small jaw), cleft palate, Legg's Calve Perthe's Disease

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ARNOLD CHIARI MALFORMATION

- ◆ Congenital elongation of the hindbrain
- ◆ Descends into the foramen magnum
- ◆ May have hydrocephalus and bony defects (ie. Basilar impression, occipitalization of C1, enlarged foramen magnum)
- ◆ May present later in life with “pseudo spondylosis”
- ◆ Neurological exam will save your *ss (buttocks)

GRISSEL'S SYNDROME

- ◆ Inflammation causing attenuation of the cranio-vertebral ligaments
- ◆ Posterior-inferior nasopharynx lacks lymph nodes
- ◆ Septic exudate in infections can flow freely through the soft tissue and into the vascular plexus surrounding odontoid
- ◆ Vascular engorgement occurs and transfers to the lateral and median AA joints
- ◆ Mechanical distension of the joints and ligaments
- ◆ Chemical hyperaemic decalcification of the bone ∴ weakens attachment of the transverse ligament (may lead to avulsion)

ARTHRITIDES

1. Rheumatoid Arthritis/ Juvenile Rheumatoid Arthritis

- ◆ Chronic inflammatory disorder characterized by synovial joint involvement
- ◆ 40% shown to have CV Region instability
- ◆ prevalence is .3 - 1.5% of the population
- ◆ women more affected than men
- ◆ inflammatory destruction of the joints via sinuvitis
- ◆ pannus erosion of the dens
- ◆ AA instability or Basilar impression of the dens into the foramen magnum
- ◆ With Occipitalization, result is usually irreversible paralysis or Death

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2. *Ankylosing Spondylitis*

- ◆ A seronegative arthropathy (associated with HLA-B27)
- ◆ Affects men > women presents in the 2nd or 3rd decade of life
- ◆ Indirect effect on the CV stability
- ◆ Progressive ankylosing of the joint interbodies along the disk
- ◆ This will fuse up to C23 – as a result of the fusion we see an increase in the motion of the AA joint (compensatory movement)
- ◆ This excessive movement becomes rapidly unstable

OTHER CONNECTIVE TISSUE DISORDERS

- ◆ Marfan's Syndrome
- ◆ Ehlers-Danlos Syndrome
- ◆ Hypermobility Syndrome

- ◆ These connective tissue disorders affect the CV region by virtue of a ligamentous laxity that develops here

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