IV CONGENITAL SPINE

KLIPPEL FLAIL SYNDROME

Prevalence 0.60%

Mainly around upper 3 vertebrae [75%]

Commonest: C2-3

Lower Cervical spine fusion may be associated with syndromes:

Fetal alcohol syndrome
Goldenhar syndrome

Triad

1. Short neck; Webb neck [Pterygium Colli]
2. Low posterior hairline
3. Restriction of neck ROM [3 or more vertebral fusion]

Other findings:

4. Torticollis

May have excessive movement in the unfused segment to compensate.
Flexion-extension better preserved than lateral bend or rotation

5. Pain is due to instability or degeneration or stenosis

6. Neurological
    1. Radicular: from osteophytes at the mobile segment
    2. Cord symptoms

7. **Types**: Henisinger: 3 pattern

Type I: C2-3 fusion with occipatalisation of C1.

Causes C1-C2 instability with age can cause spinal cord problem

Type II: Long fusion with instability Occipito-cervical spine.

Type III: Single open interphase between two fused segments
Spinal movements concentrated at the single open articulation.

There is no correlation between the fusion pattern and presence of signs or symptoms

Association

Genito-urinary system	35%
Nervous, Cardiovascular, hearing impairment	20%
Congenital Scoliosis [Hemivertebrae]	60%
Sprengel's shoulder	30%
Congenital Heart disease [Septal defects; dextro-cardiac]	14%

X ray

Number of vertebra
Fusion partial or complete
Posterior may be earlier than anterior part of the vertebra
Look for scapula and omovertebral bar
Flexion-extension: Hypermobile segment

Stenosis: Pavlov ratio <0.8
Absolute diametre <13 mm

InstabilityC1-2

5mm translation
11° angulation

TREATMENT

1. Expected to lead normal life
2. Avoid contact sports in high risk patterns of cervical spinal motion
3. Role of prophylactic fusion is not been defined
4. Arthritic pain: Treatment is symptomatic relief

BASILAR IMPRESSION

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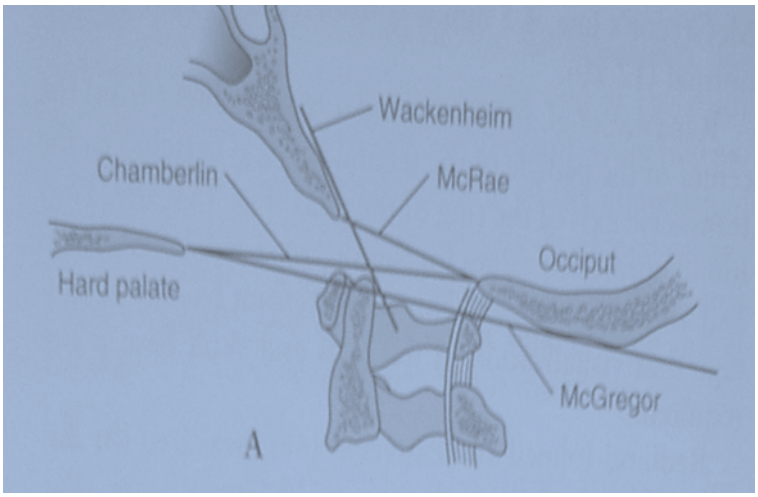
3. Role of prophylactic fusion is not been defined

X ray: McRae's line
Chamberlin's line
McGregor's lines line



Value of using these lines are unclear and can be difficult

Ideal: MRI: Tip of the odontoid above the level of foramen magnum.

Look for pressure and syringomylia

Treatment

When symptomatic:
Suboccipatal decompression + C1 & C2 laminectomy and posterior fusion from Occiput to C2

OCCIPITO-ATLANT FUSION

Known as occipitalization of the atlas

Seen in: Achondroplasia

Diastrophic Dwarfism,

SED,

Morquio syndrome

70% have neurology: usually due to a fibrous dural band or posterior element of atlas. This causes posterior column signs. May have symptoms of cerebellar herniation

Rx: Decompression and fusion of Occiput to the Atlas

OS ODONTOIDEUM

Separated upper one third of the dens
May present with instability
Aetio: ? old fracture odontoid gone on to Non-union
Congenital malformation

Increase instability with flexion-extension.

Treatment

Asymptomatic patient, only observation is required if plain X Ray does not show any instability.

Patient with Lhermitte's phenomenon with plain X ray showing instability and MRI demonstrating chronic cord lesion, there is a need C1-2 fusion

CONGENITAL SCOLIOSIS

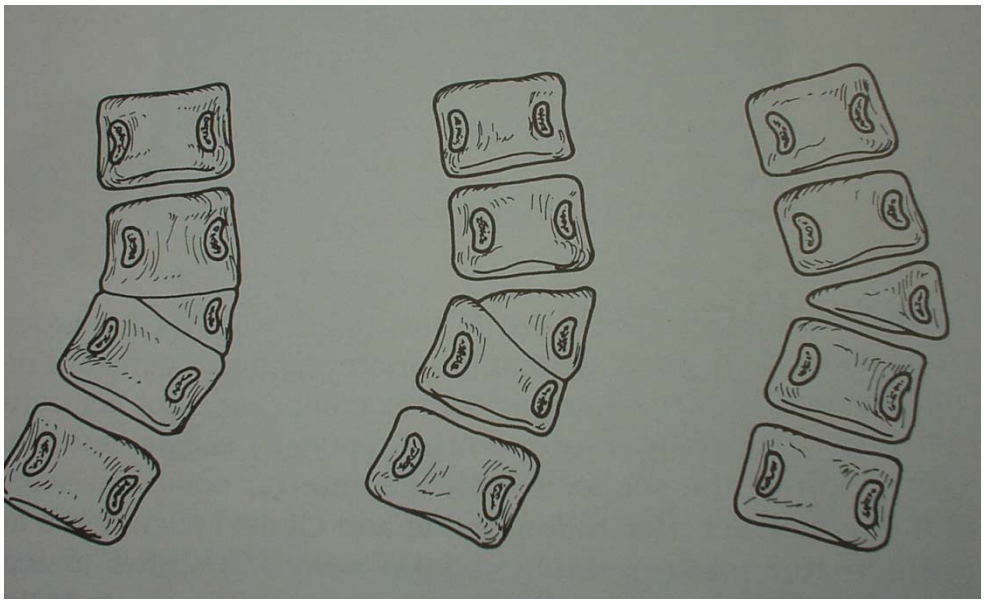
Winter's Classification

I **Failure to develop:** Hemivertebra:

a) Fully segmented [nonincarcerated] is common
b) Partial segmented
c) Non-segmented [incarcerated]

II **Failure of segmentation:** Bar

III **Combination: Wedge and Bar**



Associated anomalies: GUT, KFS 25%

Heart disorder 10%

Clinical

Deformity [progressive scoliosis]

Follow up: every 4 months

Assessment of scoliosis:

Progress of the curve: 1. Growth potential: Maximum when there is bar with Contralateral, and in nonincarcerated hemivertebra

2. Site: Thorax and thoracolumbar Hemivertebra are worse

3. Age of presentation: younger the age worse is the prognosis

Any other congenital problems [MRI]

40% intraspinal abnormalities: Spinal dysraphism [Diastometamyelia, Ductal actasia]
Syringomyelia
Low lying conus

Treatment

Bracing has little role

Casting and observe

Early surgery: in high growth potential. Ideal age is 5 years; presence progressive scoliosis; angle >40°

When surgery is considered, it requires both anterior and posterior approach

Philosophy is "Short straight spine is better than crooked long spine"

Commonly done: Excision of a fully segmented Hemivertebra and fusion through anterior and posterior approach.

CONGENITAL KYPHOSIS

1. Failure of formation is more common
2. Is likely to progress

3. Is the commonest cause for paraplegia [Neurofibromatosis is the II commonest]

4. < 4 yrs: Posterior fusion is adequate

   > 4 yrs: Posterior and anterior surgery

DIASTOMETAMYELIA

Female: Male = 8:1
2 segments: with septum by fibrous, cartilage or bone

60%: lumbar region
90% X ray shows wide interpedicular distance
40% associated with vertebral deformity like hemivertebra
10% in all congenital anomaly of the spine

Clinical

Hairy patch over the spine 75%
Calf muscle asymmetry in 50%
Cavus deformity ; unilateral in 50%
Scoliosis in 75%