CRANIOVERTEBRAL JUNCTION ANOMALIES-DIAGNOSIS AND MANAGEMENT

PRESENTER- DR ANAND V K

MODERATOR- DR PS CHANDRA

DEPARTMENT OF NEUROSURGERY AIIMS, NEW DELHI

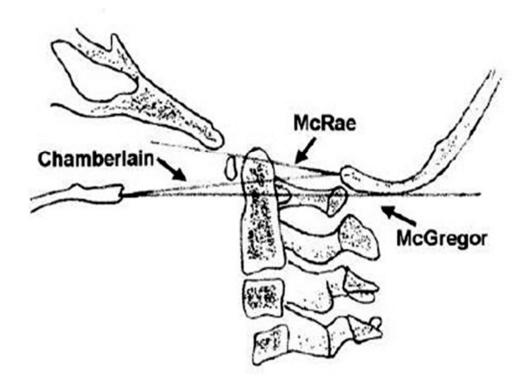
9/2/2011

CRANIOMETRY:

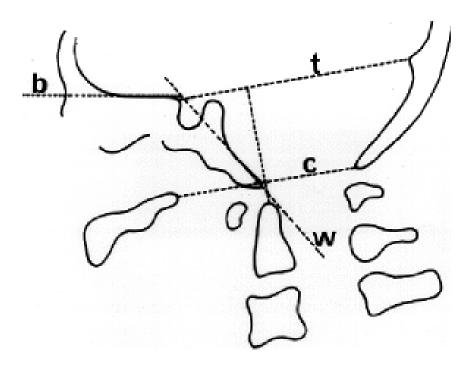
- Uses a series of lines, planes & angles to define the normal anatomic relationships of the CVJ.
- Plain Xrays,3DCT or on MRI.
- No single measurement is helpful.
- Disadvantages ;; anatomic structures and planes vary within a normal range.

LATERAL PROJECTION OF SKULL X-RAY:

- Palato–occipital (Chamberlain's line) (>5 mm)
- Palato–suboccipitalline (McGregor line) (> 7mm)
- Foramen magnum line (McRae line) (Tip of dens below this line)



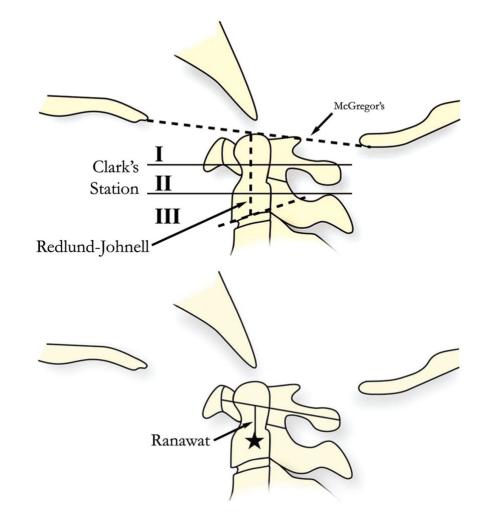
- Height of the posterior cranial fossa (Klaus Index) (<30 mm - Bl)
- Wackenhein's clival canal line (Tip of dens lies below)
- Bull's angle (Atlanto palatal angle) (< 10⁰ – normal , >13 BI)



Clark's station (BI - if ant ring of C1 is in zone II or III)

Redlund - Johnell (normal > 34 mm)

Ranawat (if < 15 mm – BI)



FRONTAL PROJECTION OF SKULL X-RAY:

- Bimastoid line (Fischgold & Metzer)
- Bidigastric line (Fischgold & Metzer)
- Condylar angle (Schmidt & Fischer)

FOR DIAGNOSIS OF PLATYBASIA:

- Basal angle (Welcher)
- Boogard's angle

OCCIPUT ANOMALIES

THE OCCIPUT :

- The basi occiput forms the lower portion of the clivus.
- The upper portion of the clivus is formed by the basisphenoid, separated from the basi occiput by the sphenooccipital synchondrosis.
- The age at which this synchondrosis fuses ranges from "after the 12 yr" – 14 yrs for girls and 16 yrs for boys.
- Most occipital anomalies are associated with decreased skull base height and basilar invagination.

CondylusTertius:

• Proatlas persists or when it fails to integrate, an ossifled remnant may be present at the distal end of the clivus, called the condylus tertius or 3rd occipital condyle.

CondylusTertius:

- This third condyle may form a joint or pseudojoint with the odontoid process or with the anterior arch of the atlas and may lead to limitation in the range of motion of the CVJ.
- There is an increased prevalence of os odontoideum associated with this abnormality.

Condylar Hypoplasia:

- In condylar hypoplasia, the occipital condyles are underdeveloped and have a flattened appearance, leading to BI (violation of the Chamberlain line) and widening of the atlanto occipital joint axis angle.
- The lateral masses of the atlas may be fused to the hypoplastic condyles, further accentuating the BI.
- Condylar hypoplasia limits, or may even abolish, movements at the A-O joint.

Atlanto-occipital Assimilation / Occipitalization of Atlas:

- The failure of segmentation between the skull and first cervical vertebra results in assimilation of the atlas.
- Incidence 0.08 to 3 %.
- India 40 to 50 % are associated among CVJ anomalies.
- The assimilation may be complete or partial.
- It invariably results in basilar invagination.
- Wackenheim clivus baseline may be normal, but the clivus-canal angle may be decreased.

TOPOGRAPHIC FORMS (WACKENHEIM):

- Type I: Occipitalization (generally subtotal) associated with BI.
- Type II: Occipitalization (generally subtotal) associated with BI & fusion of axis & 3rdcervical vertebrae.
- Type III: Total or subtotal occipitalization with BI & maldevelopment of the transverse ligament.
- Type III may be associated with various malformations like C2-C3 fusion, hemivertebra, dens aplasia, tertiary condyle, etc.

Contd...

- The neurological symptoms are not caused by occipitalization proper but rather by the fact that in the absence of a free atlas, TL fails to develop which causes posterior displacement of axis & compression of the spinal cord.
- Instability b/w atlas & axis is reducible in patients <15 yr's but after that irreducible state occurs.
- Its combination with segmentation failure of 2nd& 3rdcervical vertebrae results in gradual loosening of the atlanto-dental joint with progressive atlanto-axial luxation in children in about 50% of cases.

ATLAS :

- With the exception of the various atlanto occipital assimilations, most atlas anomalies, when isolated, produce no abnormal CVJ relationships and are not associated with basilar invagination.
- The vast majority of anomalies consist of various arch clefts, aplasias, and hypoplasias.
- Arch anomalies are frequently mistaken for fractures in the evaluation of plain radiographs of patients with a history of cervical spine trauma.

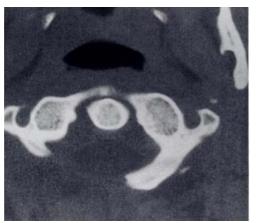
<u>PONTICULUS POSTICUS / KIMMERLE'S</u> <u>DEFORMITY :</u>

- It is a bony ridge projecting posteriorly from the articular edge of the atlas superior articular facet.
- The bony projection may be only a few mm long or may elongate to unite with the adjacent neural arch of the atlas to produce an "ARCUATE CANAL" through which the vertebral artery passes.
- This is due to ossification of a portion of the oblique A-O ligament.

Ponticulus posticus



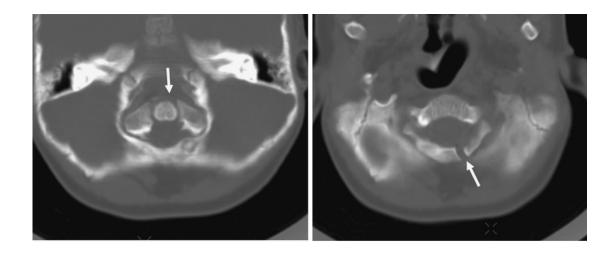
Posterior Arch Anomalies (MC atlas anomaly) :



- Total or partial aplasia of the posterior atlas arch is rare.
- Although absence of the posterior arch, when isolated, is usually asymptomatic, but may be associated with anterior atlantoaxial subluxation.
- Bilateral atlantoaxial subluxation may be associated with both total and partial aplasias, simulating the Jefferson fracture.

Contd...

- Clefts of the atlas arches are much more common.
- Posterior rachischis is, most common, is observed in 4% of adults.
- The majority of posterior atlas clefts (97%) are midline, whereas lateral clefts, through the sulcus of the vertebral artery, account for the remaining 3%.
- Posterior arch rachischis may be superimposed on the odontoid process or the axis body on the openmouth odontoid view, simulating a fracture.



SPLIT ATLAS:

- In contrast to posterior arch rachischis, anterior arch rachischis quite rare (0. 1 %).
- Typically encountered in association with posterior rachischis -"*split atlas*".
- Normally, on a lateral radiograph, the anterior arch of the atlas appears crescentic or half-moon-shaped, with dens cortical bone surrounding the medullary cavity and a well-defined predental space.
- In anterior arch rachischis, the anterior arch appears fat or plump and rounded in configuration, appearing to "overlap" the odontoid process (making identification of the predental space impossible); the arch may have un sharp, duplicated anterior margins

CONGENITAL ODONTOID ANOMALIES OR DYSPLASIAS

- Type I : Os odontoideum
- Type II : Ossiculum terminale
- Type III : Agenesis of odontoid base
- Type IV : Agenesis of apical segment
- Type V : Agenesis of odontoid

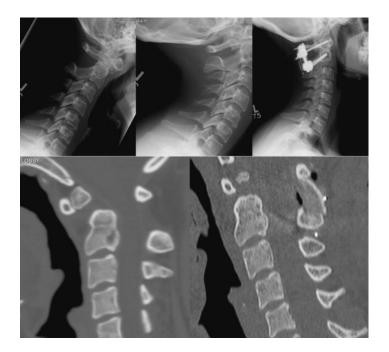
OS ODONTOIDEUM

- Giacomini (1886),
- independent osseous structure lying cephalad to the axis body in the location of the odontoid process.
- The anterior arch of C1 rounded, posterior arch hypoplastic.
- Congenital / Traumatic (current Hypothesis) / vascular.
- Orthotopic / Dystopic (unstable)
- Associated with connective tissue disorders.

Os Odontoideum : Etiology and surgical Management ; ARVIN ET AL ; Neurosurgery / vol 66/ no 3/ march 2010 suppl

Contd...

- Reducible on flexion, dorsal compression of the cord occurs but on extension ventral compression occurs secondary to increased angulation anteriorly.
- Irreducible due to displacement of TL ventral to the ossicle.



- C/F Asymptomatic
 - Neck pain --- C2 may be only

symptom

- myelopathy.
- VBI
- D/D Ossiculum terminale Persistence

OS ODONTOIDEUM

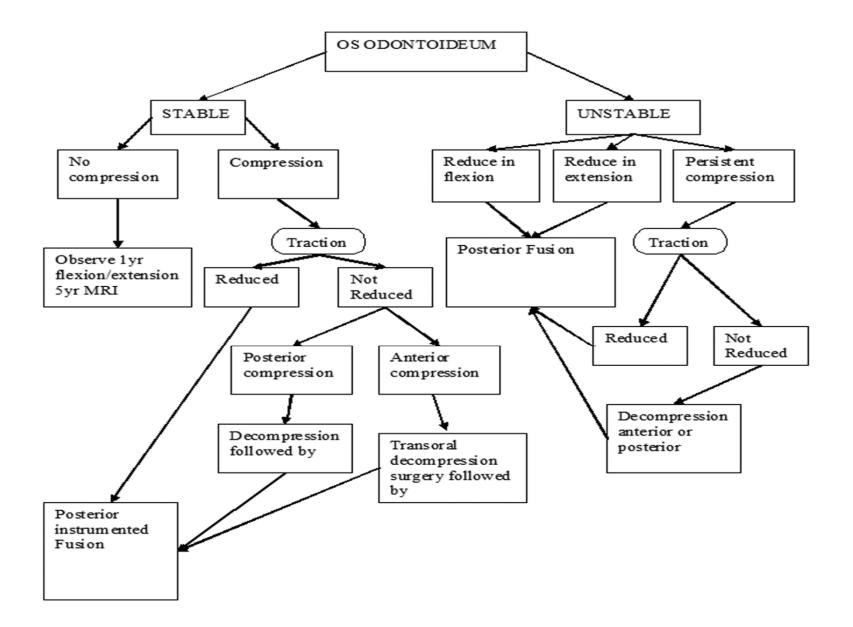
Treatment:

□ Use of traction --- Controversial.

Surgical Techniques:

Goal --- Remove compression at FM

--- Stabilize spine in relation to subaxial spine.



Suggested algorithm for the management of a patient with os odontoideum. MRI, magnetic resonance imaging.

<u>Persistent</u> OssiculumTerminale



- Also called Bergman ossicle, results from failure of fusion of the terminal ossicle to the remainder of the odontoid process.
- The fusion typically is accomplished by 12 years of age.
- Bergman ossicle may be confused with a type 1 odontoid fracture (avulsion of the terminal ossicle), and absolute differentiation between the two diagnoses may be difficult.
- Whether traumatic or congenital in origin, this anomaly is stable when isolated and of relatively little clinical significance.
- The odontoid process is usually normal in height.

Odontoid Aplasia:

- Total aplasia of the odontoid process is extremely rare.
- A true aplasia is associated with an excavation defect into the body of axis.
- may simulate os odontoideum, as the os fragment may be perfectly projected over the atlas arch on the open mouth odontoid view.

KLIPPEL FEIL SYNDROME

- Described first by Klippel and Feil in 1912.
- Etiology is unknown.
- Segmentation failure of cervical somites during the third and eighth weeks of gestation.
- Classic triad (50%) --- Low posterior hair line, short neck and limited neck movements.

TYPES:

- Type 1: Cervical spine fusion in which elements of many vertebrae are incorporated into a single block.
- Type 2: Cervical spine fusion in which there is failure of complete segmentation at only one or two cervical levels and may include an occipito – atlantal fusion.
- Type 3: Type 1 or type 2 fusion with co-existing segmentation errors in the lower dorsal or lumbar spine.

KLIPPEL FEIL SYNDROME

ASSOCIATED CONDITIONS:

- Scoliosis- Up to 60% have >15 degrees curve.
- Genito- urinary- up to 65%.Most common is absence of kidney.
- Sprengel's deformity- approx. 35%
- Cardio pulmonary- 5-15%, most commonly V.S.D.
- Deafness-30%, all types, MC mixed.
- Sykinesis Mirror motions have been described in up to 20% of patients under the age of 5.
- Cranio- cervical abnormalities (25%) -Includes C1-C2 hypermobility and instability, BI, Chiari I malformation, diastematomyelia, & syringomyelia.

KLIPPEL FEIL SYNDROME

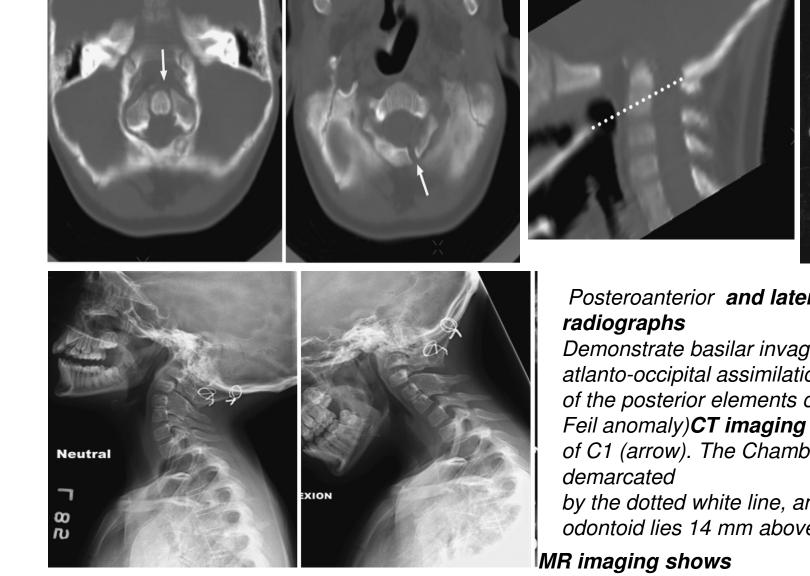
<u>SYMPTOMS:</u>

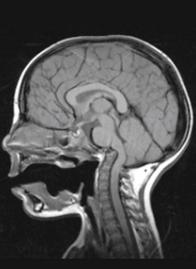
- Due to the hypermobility occurring at the open segments, can lead to either frank instability or osteoarthritis.
- Mechanical symptoms due to joint irritation.
- Neurologic symptoms due to root irritation or spinal cord compression.

<u>MANAGEMENT :</u>

• Usually conservative, rarely surgery required.

The CVJ and its Abnormalities by John C. VanGilder / Arnold H. Menezes / Kenneth D.Dolan ; University of Iowa School of Medicine.





Posteroanterior and lateral cervical

Demonstrate basilar invagination, partial atlanto-occipital assimilation, and fusion of the posterior elements of C4 to 5 (Klippel-Feil anomaly)CT imaging shows unfused ring of C1 (arrow). The Chamberlain line is

by the dotted white line, and the tip of the odontoid lies 14 mm above this line.

Severe BI follow up x -ray cervical spine

Smith and Menezes ; Basilar Invagination : Neurosurgery vol 66 / no 3/ march 2010 suppl

- The floor of the skull is indented by the upper cervical spine, & hence the tip of odontoid is more cephalad protruding into the FM.
- Primary invagination developmental, MC
- Secondary invagination -- Acquired.
- Primary BI--- associated with occipitoatlantal fusion, hypoplasia of the atlas, a bifid posterior arch of the atlas, odontoid anomalies.
- 25 35% -- ACM, Syringomyelia, Hydrocephalous
- VA anomalies common

Topographic types of BI :

- Anterior BI : hypoplasia of the basilar process of the occipital bone.
- Posterior BI: posterior margin of the FM is invaginated.
- BI in the lateral condylar area.
- Unilateral BI.
- BI of the occipital condyles (Paramedian BI) Condylar hypoplasia
- Generalized BI

- <u>SIGNS / SYMPTOMS</u>: usually occur in 2nd or 3rd decade.
- Short neck(78%), torticollis (68%), low hair line (48%), Webbed neck (47%) are local findings
- s/s of associated ACM (cerebellar & vestibular disturbances) & syringomyelia (25 to 35%).
- Motor & sensory disturbances (85%).
- Lower cranial nerves involvement
- Headache & pain in the nape of neck (greater occipital N)
- s/s of raised ICP (HCP) due to posterior encroachment which causes blockage of aqueduct of sylvius.
- Compression of cerebellum & vestibular apparatus leading to vertical or lateral nystagmus (65%) (not due to direct pressure from post rim of FM but rather due to a thickened band of dura).
- Vertebral artery insufficiency s/s.

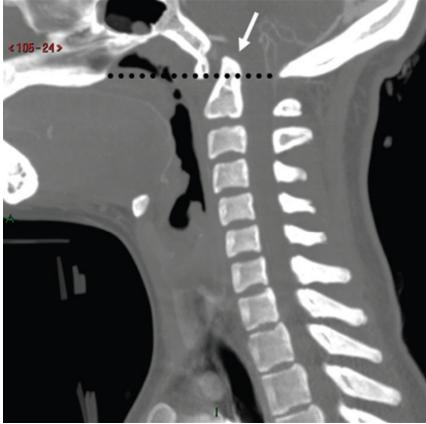
IMAGING

Imaging Parameters:

 Should be suspected if C1C2 facet complex can not be visualized in open mouth AP view of CVJ X-ray.

Lateral Xray:

- Chamberlain line 5 mm.
- Mc Gregor line 7 mm.
- Mc Rae line Tip should be below .
 --- FM < 19 mm.
- MRI --- Soft tissue , ligaments ,cord signal changes, severity of BI



Surgical treatment : Indications

- All symptomatic patients.
- Subset of neurologically intact patients with mild BI and lack of significant compression may be followed up.
- If Reducible --- posterior decompression with fusion.
- Irreducible --- Anterior decompression + Posterior fusion.

CVJ ANOMALIES IN DOWN SYNDROME

- First described in 1965 by Tishler & Martel.
- Characterized by increased ligamentous laxity and abnormal joint and bony anatomy (abnormal curved shape of Occipital condyle and C1 articular facet).
- Incidence of radiologic AA instability -- 7-40%,
 < 1% of patients are symptomatic.
- Bony anomalies involving the occipital condyle, C1 ring, and Os odontoideum (6%) also increased in Down syndrome.
- Dense not secured against ant arch of C1 during flexion / Extension. May also have rotatory instability.

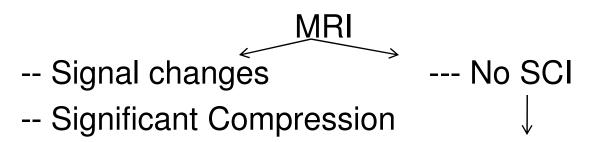


Pre op flexion / extension X-ray and Dynamic MRI (T2WI , Sag) images in Down syndrome

CVJ ANOMALIES IN DOWN SYNDROME

Radiological Screening & Recommendations (AAP -2007)

- Os odontoideum --- Sx.
- Canal width < 14 mm
 with or without ADI > 5 mm



- -- Follow up 1 yr
- -- Restriction of high risk activities
- Symptomatic compressions surgery.

CVJ ANOMALIES IN DOWN SYNDROME

SURGICAL TREATMENT :

- In most cases Posterior instrumentation + Fusion. Transoral decompression – if irreducible Ventral compression.
- Fusion rate 58 95% (Screw based fixation)
 Wire based fixation 40% (? Resorbed bone graft)

Hakinson and Anderson et al : CVJ abnormalies in Down syndrome : Neurosurgery/vol 66/ no 3/ march 2010 suppl

GREENBERG'S CLASSIFICATION;

- Incompetence of the odontoid process-
- Incompetence of the Transverse ligament

WADIA CLASSIFICATION :

- Group I: AAD with occipitalization of atlas & fusion of C2 & C3.
- Group II: odontoid in competence due to its maldevelopment with no occipitalization of atlas.
- Group III: odontoid dislocation but no maldevelopment of dens or occipitalization of atlas.
- 57% of all CVJ anomalies.
- 8.3% of all causes of cervical compression

- On the open mouth odontoid view, the combined spread of the lateral masses of C1 on C2 should not exceed 6.9 mm.
- > 6.9 mm -- rupture of the transverse ligament.
- An atlantoaxial distance greater than 4-5 mm, as demonstrated by lateral radiographs, is indicative of AAI.



- Posterior atlantodental interval (PADI) measured from the posterior border of the dens to the anterior border of the posterior tubercle.
- This index may be more important because it more directly assesses the spinal canal width.
- Normal range 19 –32 mm in male & 19 –30mm in females.
- Below 19mm, neurological manifestations occur.



- Type I injuries (stable subluxations) Hard Collar.
- Type II injuries may be potentially unstable.
- Type III and IV rotatory displacements that are unstable are treated surgically with a reduction and C1-2 fusion.
- The techniques of fusion vary from sublaminar wiring techniques like Brooks or Gallie, Halifax clamp, or transarticular screw of Magerl.

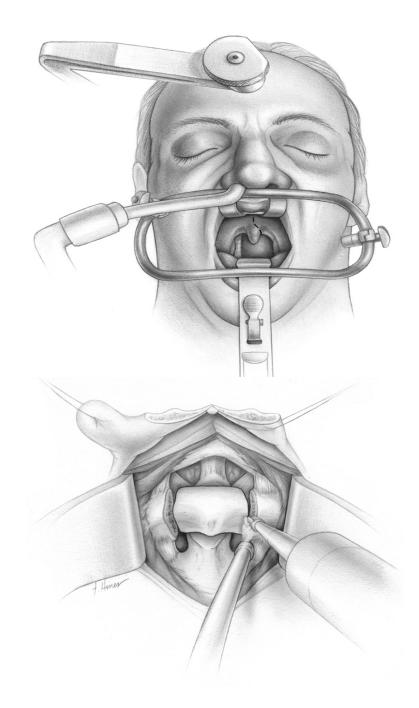
SURGICAL APPROACHES FOR VENTRAL DECOMPRESSION

- Anterior :
 - Standard transoral approach.
 - Extended transoral approaches.
 - --- Transoral mandibulotomy
 - --- Transoral mandibulotomy + Palatotomy
 - --- Transoral mandibulotomy + glossotomy
 - --- Transoral + Lefort I Maxillotomy.
 - Endoscopic transoral approach
- Lateral extrapharyngeal approach

TRANS ORAL APPROACHES

- 1st by kanavel (1919)
- Fang and Ong (1962)
- Arnold Menezes (1977)
- Interdental opening -- > 25mm
- Why ?
 - --- Performed in midline avascular plane
 - --- most direct route to osseous and soft tissue abnormalities ventral to brainstem & CVJ.
 - --- ICA; Masticatory muscles, TM Joint VII N, Vestibulocochlear apparatus are avoided

- Important to identify C1 tubercle.
- 15 mm wide trough is created in the arch of C1.
- Along with odontoid, Alar, apical, transverse ligaments should be severed.
- Remove all the granulation tissue till pulsatile dura clearly visible.
- Inadvertant dural puncture Sealed immediately.



- Standard approach :
 - 1) Lower clivus to C2C3 interface.
 - 2) No uniform consensus regarding extent of exposure.
 - 3) palatotomy increases exposure to mid clivus.
- Mandibulotomy ;
 - --- shortens the operative distance.
 - --- Provides wide angle.
 - --- mid clivus to C2C3 interface
- Mandibulo glossotomy ;
 - --- Further reduces the distance
 - --- Upper 1/3 clivus to C3-C4 interface.
- Mandibulo glossotomy + Palatotomy ;
 - --- extends rostral exposure to spenoid sinus.
 - --- At the cost of velopharyneleal insufficiency (30%) dysphagia, nasal regurgitation hypernasal voice.

COMPLICATIONS

- Early
- -- bleeding
- -- soft tissue swelling
- -- infection
- -- CSF leak
- -- wound breakdown
- -- dislocated mandible
- -- nausea, vomiting, aspiration
- -- blocked eustachian tube

<u>Late</u>

- -- velopharyngeal insufficiency
- -- nasal regurgitation
- -- nasal speech
- -- poor mouth opening
- -- dental malocclusion

Intermediate

- -- delayed arterial bleeding
- -- wound break down
- -- wound / chest infection
- -- CSF leak
- -- velopharyngeal insufficiency
- -- nasal voice
- -- nasal regurgitation

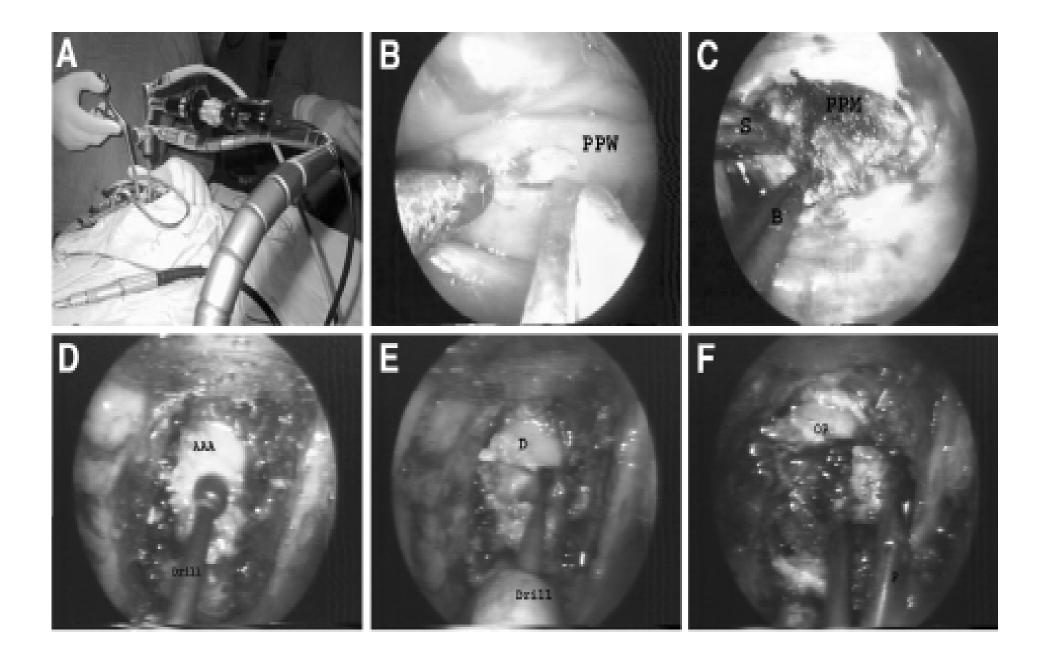
ENDOSCOPIC APPROACH

- 0° right angled endoscope for illumination.
- Endoscope brought into the left side of oral cavity and fixed with table mounted holding device.
- Incision made for clivus to C2.
- Lateral limit of exposure kept at 15 mm on each side.
- Under endoscopic view ant arch of C1 drilled to expose dens.
- Dens drilling conducted from base.

BENEFITS OF ENDOSCOPE

- Viewing angle endoscopes obviate the need for soft palate split.
- Difficulty in viewing through the microscope at retrouvular region.
- Can effectively preserve Velopharyngeal function.

Endoscopic transoral surgery for CVJ anomalies; M. Husain et al ; J Neurosurgery : Spine/ vol 5/ oct 2006



C1 - C2 FIXATION

- First described by Hadra in 1891
- Modified by Cone in 1937
- Three basic cable wire fixation
 - Gallie
 - Brooks
 - Sonntag modification of gallies technique.
- Indicated in AAD with intact C1C2 laminae
- Contraindication
 - Absent posterior element
 - Severe osteoporosis
 - Narrow canal diameter
- Fusion rates increase when external orthosis is used
- Quadriparesis can occur upto 5 to 7 % of cases
- Breakage of wire might occur

GALLIE

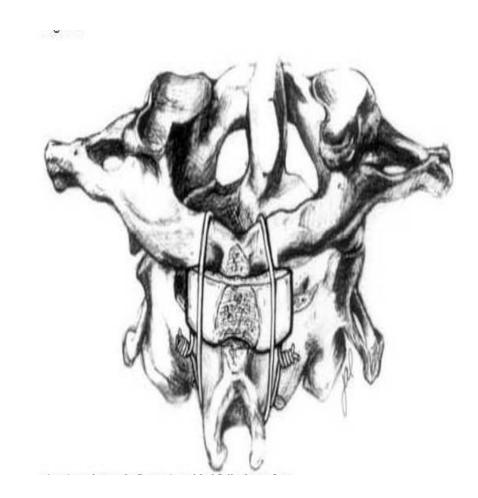
- 20 gauge wire under C1 arch
- Inferior part notched over spinous process of C2
- H shaped graft placed in between decorticated C1 and C2 arch
- Notch inferiorly in the midline
- Offer little rotational or extension stability
- High rate of non union

BROOKS

- Superior to gallie in biomechanical studies esp. rotation
- Caries risk of passing additional wire beneath two lamina
- Two rectangular bone graft fashioned to wedge between C1 and C2

SONNTAG DICKMAN

- One bicortical graft wedged
 between C1 and C2
- Single sublaminar wire
 passed under C1
- Wire looped below C2 spinous process trapping the graft between C1 C2
- Avoids passing second sublaminar wire



LAMINAR CLAMPS

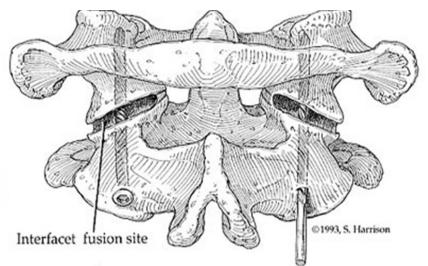
- Offers posterior laminar fixation without passing sublaminar wire
- Upper clamp placed above lamina of C1
- Inferior clamp placed below C2
- Sequential tightening of clamps to ensure engagement of laminae
- Interlaminar graft may be used
- Associated with implant slippage
- High pseudoarthrosis rate
- C1 ring fracture

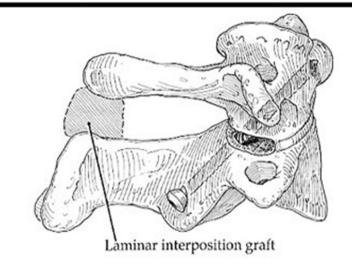
TRANSARTICULAR SCREW FIXATION

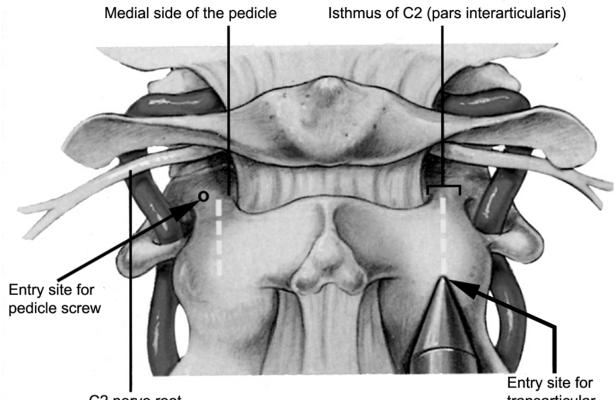
- Used when posterior arches incompetent
- Superior to wiring methods in fusion rates
- Severe osteopenic patients may require fixation through articular facets
- Imperative to delineate vertebral arteries course
- not feasible if vertebral arteries are too medial
- Provides absolutely no movement in the region
- Procedure is technically difficult
- Spinal cord injury, hypoglossal injury, vertebral artery injury 4 8%.
- Magerl technique provides stiffest stabilization with least amount of rotation and lateral bending
- Provides three point fixation
- Follows most closely biomechanical rules on internal fixation
- Fusion rates between 85% to 98% and well tolerated in elderly

POSTERIOR TRANSARTICULAR SCREW

- Point of entry 2mm up ,2mm in from the medial ½ of inferior facet of C2.
- Transarticular screw placed through pars interarticularis of C2 penetrating C1 C2 facet into lateral masses of C1
- Screw directed 25^o towards midline and 25^o cranial
- Three point fixation can be obtained









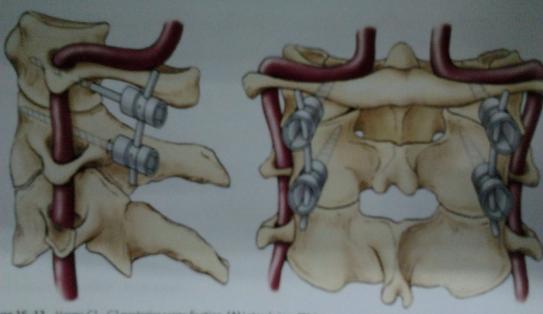
困っ

C2 nerve root

Entry site for transarticular screw

LATERAL MASS SCREW FIXATION

- Jurgen Harms technique of C1 C2 fixation
- C1 lateral mass screw and C2 pedicle screw with rod reconstruction
- Reduction of C1 C2 can be achieved using rod as a lever arm
- Can be used in patients with compromised posterior element
- Easier than placement of transarticular screws



C1 LATERAL MASS WITH C2 PEDICLE SCREW WITH PLATE FIXATION

- Provides immediate stability
- Achieved 100% fusion rate
- Anatomic alignment not necessary
- Technically demanding
- Distraction of C1 C2 joint space necessary.
- C2 nerve root mobilized
- 3.5 mm poly axial screws (26 34 mm) are used.



Goel A, Laheri Y. Place and screw fixation for atlanto – axial subluation, Acta Neurochir (Wien). 1994; 129 (1-2):47-53

OCCIPITO CERVICAL STABILIZATION

- Indicated in pt with AAD occipitalised hypoplastic or bifid atlas
- Posterior decompression of foramen magnum and C1 arch required especially Arnold chiari
- Wire and bone graft fusion by artificial arch of atlas
- Bony ridge of occipital bone and occipitalized atlas created
- Artificial arch and C2 decorticated and one midline and two lateral notch formed
- Wire passed through the notch
- Graft of iliac crest wedged between C2 and artificial arch and wire is tightened
- Rarely used now.

OCCIPITO CERVICAL FUSION

- Contoured rods
- C1 arch removed
- Rod is placed over occipital bone and c2 and c3
- Rod fixed over occipital bone by three burr holes and wires passed through it
- Absolute bony contract is required to prevent vertical movements

OCCIPITO CERVICAL FUSION

- Currently MC used suboccipital fixation method is bicortical screw fixation into midline suboccipital keel and paramedian cranium.
- 4.5/6 mm for lateral screws, 4.5/8 –12 mm for midline screws.

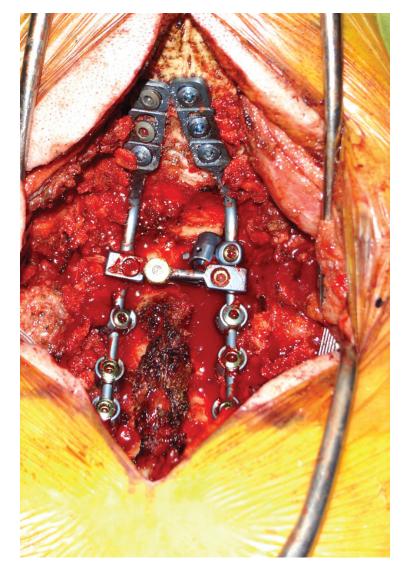
1-piece "inverted U" or "inverted Y"-shaped rod/plate with occipital and cervical wiring and/or screws

Modular (more than 1 piece) occipital plate-cervical rod constructs with occipital and cervical screws

Occipital buttons connected to cervical rods

Condylar polyaxial screws connected to cervical rods

- Cervical fixation
 - Lateral mass screws
 - Laminar screws
 - Transarticular screws
 - or any of combinations.
- Provides immediate stabilization
- Screw and plate rod fixation instrumentation allows surgeon to determine the optimal screw fixation point and to perform deformity correction
- Highest osseous fusion rate (94-97%).



Nuances of Occipital fixation – P. V Mummaneni et al Neurosurgery/ vol 66/ no 3/ march 2010 suppl.

