PEDIATRIC CERVICAL SPINE DEFORMITY

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DEVELOPMENT

- EACH VERTEBRAE DEVELOP FROM THE CAUDAL AND CRANIAL ½ OF 2 SCLEROTOMES
  - C1 and C2 primitive centrum fuse to form odontoid process

- ATLAS
  - Body ossifies at 6-24 mos
  - Arch closure (final canal diameter) at 6-7 yo
    - Further growth by periosteal appositional growth only (external, no canal change)
DEVELOPMENT

• AXIS
  – ODONTOID
    • 2 PRIMARY OSSIFICATION CENTERS
      – Coalesce by 3 mos
      – Separated from C2 by dentocentral synchondrosis
        🌟 Gradually closes btwn 3-6yo
    • TIP BECOMES AN APOPHYSIS
      – Chondrum terminale
      – Begins ossifying 5-8yo
      – Fuses at 10-13yo
  – NEURAL ARCHES CLOSED BY 6-7 YRS
    • Similar to atlas, no increase in canal size after this point
DEVELOPMENT

• C 3-7
  - 3 OSSIF. CTNS @ BIRTH
    • 1 BODY, 2 ARCHES
    • POST SYNCHONDROSIS CLOSES @ 2 YRS
  - FACETS START
    HORIZONTAL, BECOME VERTICAL WITH GROWTH
DEVELOPMENT

• C 3-7
  – BODY GROWTH
    • VERTICAL – ENCHONDRAL OSSIFICATION
    • CIRCUMFERENTIAL – PERIOSTEAL APPOSITION
RADIOGRAPHIC PARAMETERS

- WHAT IS PATHOLOGIC IN AN ADULT CAN BE NORMAL IN A GROWING CHILD
  - ADI
  - SAC
  - PSEUDOSUBLUXATION C 1-2
  - OS-ODONTOIDUM
  - GROWTH CENTERS ARE NOT FRACTURES
RADIOGRAPHIC PARAMETERS

• ADI (ATLANTO-DENS INTERVAL)
  - Measure on lateral flex/ext films,
    voluntary motion in awake patient
  
• ANT ASPECT OF DENS TO THE POST ASPECT OF THE ANT RING OF THE ATLAS ON BOTH FILMS
  - NL: < 5mm kids,
    <3mm adults
RADIOGRAPHIC PARAMETERS

• Anterior arch of atlas can override odontoid on extension in 20% of kids

• Why ADI increase in kids?
  – ↑ ligamentous laxity
  – ↑ cartilage component of dens and atlas
RADIOGRAPHIC PARAMETERS

• SAC (SPACE AVAILABLE FOR THE CORD)
  - POST ASPECT OF DENS TO ANT ASPECT OF POST RING OF ATLAS
  - >13 mm in adults and teens
  - Need at least the diameter of the odontoid available
RADIOGRAPHIC PARAMETERS

- **SAC**
  - Steel’s rule of thirds
    - 1/3 cord
    - 1/3 odontoid
    - 1/3 space available (“safe zone”)
  - ATTENUATION OF TRANSVERSE ATLANTAL LIG LEAVES ONLY THE ALAR LIG (i.e. TRISOMY)
    - ALAR LIG ALONE CANNOT PROTECT FROM SCI WITH EVEN MILD TRAUMA
COMMON NORMAL VARIANTS

• Absence of cervical lordosis
  - Mimics splinting of injury
• Pseudosubluxation
• C1 multiple ossification centers/ spina bifida
  - Can mimic fx
  - Look for smooth cortical margins
  - Lack hematoma on CT
• Spina bifida occulta
• C2 dentocentral synchondrosis
  - CLOSES BY 11 YEARS
• Anterior wedging of C3 seen in 7%
PSEUDOSUBLUXATION

- **ANT DISPLACE OF C2 ON C3**
  - C 3-4 less common
  - 9% of kids 1-7 yo
  - Posterior line of Swischuk
    - Line from ant aspect of C1 posterior arch to same on C3
    - Should be within 1 mm of same of C2
    - >2 mm = pathologic
  - **CAUSES**
    - Horizontal facets
      - Esp. in upper
    - ↑ RELATIVE HEAD SIZE
    - ↑ LIG LAXITY
PSEUDOSUBLUXATION

• **ANT DISPLACE OF C2 ON C3**
  - **CAUSES**
    - Horizontal facets
      - Esp. in upper C spine
      - Change from 30deg to 70deg during growth
    - Large relative head size
    - General ligamentous laxity
  - **Treatment**
    - Do nothing
OS ODONTOIIDUM

• TIP OF ODONTOID IS DIVIDED
  - Apical segment lacks basilar support

• VERY RARE

• X-RAY – oval ossicle, smooth margins

• CAUSES ?
  - Old fx non-union
    • MRI’s have shown cord changes c/w trauma
  - AVN
  - Congenital anomaly
OS ODONTOIDUM

• SYMPTOMS
  - NECK PAIN
  - VERT ART OCLUSION (C1-2 MOTION)
    • SYNCOPE, VERTIGO, N/V, VISUAL DEFECITS
  - NEURO SXS (RARE)
    • Posterior translation of os into cord
    • Transient paresis, myelopathy, paralysis
    • SUDDEN DEATH
OS ODONTOIDIUM

• TREATMENT

  – SURGERY (C1-2 PSA, INST, HALO)
    • ADI > 10 mm
    • SAC ≤ 13mm
    • NEUROLOGIC SX
    • PERSISTANT PAIN
    • PROGRESSIVE INSTABILITY
TORTICOLLIS

- Combined head tilt and rotatory deformity
- Indicates C1-2 problem
  - 50% rotation in C-spine at this joint
- Large differential diagnosis
  - Osseous vs. nonosseous
TORTI COLLIS

- DIFFERENTIAL DIAGNOSES
  - MUSCULAR (82%)
  - ATLANTO-AXIAL ROTATORY SUBLUXATION
  - CNS LESION
  - BIRTH TRAUMA
  - CONGENITAL SPINE DEFORMITY
    - KLIPPEL-FEIL
    - OCCIPITO-CERVICAL SYNOSTOSIS
    - GOLDENHAR SYND
    - HEMI ATLAS
  - BASILAR IMPRESSION
  - ODONTOID ANOMALY (OS ODONTOID DI UM)
CONGENITAL MUSCULAR TORTICOLLIS

- 82% OF ALL TORTICOLLIS
  - 75% right sided
  - 8-20% also have DDH
- CONGENITAL CONSTRICITION OF SCM
- HEAD TILT WITH ROTATION OPPOSITE TILT
- FAMILIAR COMPONENT
CONGENITAL MUSCULAR TORTI COLLIS

- CAUSE UNKNOWN
  - INTRAUTERINE SCM COMPARTMENT SYNDROME FROM NECK COMPRESSION
    - SCM VENUS OCCLUSION ON HISTOPATHOLOGY
    - MYOFIBROSIS → CONTRACTION
  - NEUROLOGIC
    - Spinal accessory N. injury
  - FETAL POSITION
  - EMBRYOLOGIC
  - BIRTH TRAUMA
CONGENITAL MUSCULAR TORTICOLLIS

• PLAGYCEPHALY
  - Flattening of head on side of contracture
  - Due to sleeping position (prone in U.S.)
  - Untreated: eye/ear levels become unequal

• X- RAYS
  - Always normal in congenital muscular torticollis
  - Check hips

• RARELY A TREATABLE NEUROLOGIC CAUSE
  • SYRINX, SPINAL CORD TUMOR, CHIARI, POST FOSSA TUMOR, OCULAR PATHOLOGY (involuntary head tilt)
CONGENITAL MUSCULAR TORTICOLLIS

• TREATMENT
  - 90% RESOLVE WITHOUT SURGERY
    • STRETCHING, PT
    • Crib toy modification
  - After 1yo, stretching usually unsuccessful
  - SURGERY: GOOD RESULTS UP TO 12 YO
    • UNIPOLAR RELEASE
    • BI-POLAR RELEASE
      - Z-LENGTHENING OF STERNAL INSERTION MAINTAINS NECK CONTOUR
    • MID-SCM TRANSECTION
  - MINERVA BRACE FOR 6-12 WEEKS
CONGENITAL MUSCULAR TORTICOLLIS

• TREATMENT
  - FACIAL ASSYMMETRY CORRECTION RELATIVE TO GROWTH REMAINING AT TIME OF CORRECTION (REMODELING)
  - BEST SURGERY TIME BETWEEN 1 AND 4YO
ATLANTO-AXIAL ROTARY SUBLUXATION

• COMMON PROBLEM
• RANGE- MILD SUBLUX TO COMPLETE DISLOCATION
• X-RAY: difficult to assess
  • LAT MASS OF C1 Shifts anteri orly, appears wider than the narrower, more post displaced opposite lat mass
  • posteri or arches don’t superimpose due to head tilt
  • can be NL child with rotated head
  • CT with left and right rotation will demonstrate it
    - C1/O Donto id relationship is constant in a fixed deformity
ATLANTO-AXIAL ROTARY SUBLUXATION

• 4 TYPES:
  - I) ROTATORY DISPLACEMENT, NO ANTERIOR SHIFT (most common)
    • RESOLVES SPONTANEOUSLY
  - II) ROT DISPLACEMENT, WITH < 5mm ANTERIOR SHIFT
    • NOTED ON FLEX/EXT X-RAYS (ADI)
  - III) ROT DISPLACEMENT, > 5mm ANTERIOR SHIFT (very rare)
    • HIGH RISK OF PARALYSIS/DEATH
  - IV) ROT DISPLACEMENT, POST SHIFT (very rare)
    • HIGH RISK OF PARALYSIS/DEATH
ATLANTO-AXIAL ROTARY SUBLUXATION

• ETIOLOGY
  – MINOR TRAUMA
    • MOST COMMON
  – FRACTURE
  – FOLLOWING ENT SURGERY
  – FOLLOWING URI
ATLANTO-AXIAL ROTARY SUBLUXATION

• PRESENTATION
  - ACUTE TORTICOLLIS
  - PAIN WITH NECK ROM
    • Long SCM painful due to resisting deformity
  - PLAGYCEPHALY SEEN IN LONG-STANDING CASES
ATLANTO-AXIAL ROTARY SUBLUXATION

• GRISEL SYNDROME
  - SPONTANEOUS ATLANTO-AXIAL SUBLUX FOLLOWING URI, OR ENT SURG (T/A)
  - DIRECT CONNECTION BETWEEN PERIDONTAL VENOUS PLEXUS AND SUBOCcipital Epidural Sinuses
    • TRANSPORTS SEPTIC EXUDATES CAUSING ATLANTO-AXIAL HYPEREMIA
      - TEMPORARY LIGAMENTOUS LAXITY
      - SUBLUXATION
  - CHILDREN’S FACETS MORE HORIZONTAL
    • FACETS HAVE MENISCUS-LIKE SYNOVIAL POCKETS → CAN BE TRAPPED
  - USUALLY RESOLVES SPONTANEOUSLY
ATLANTO-AXIAL ROTARY SUBLUXATION

• TREATMENT
  – < 1 WEEK OF SYMPTOMS
    • SOFT COLLAR FOR COMFORT, REST
    • IF NO REDUCTION: HALTER TRACTION, VALIUM
  – 1 – 4 WEEKS OF SYMPTOMS
    • ADMIT, HALTER TX, MAY NEED HALO TX
    • CONFIRM REDUCTION ON DYNAMIC CT
    • IF NO ANT DISPLACEMENT, SOFT COLLAR 1-2 WK
    • IF ANT DISPLACEMENT, BRACE TO IMMOBILIZE FOR 6 WEEKS (allow ligaments to heal)
ATLANTO-AXIAL ROTARY SUBLUXATION

- > 1 MTH
  - HALO TRACTION FOR THREE WEEKS
  - MANY WILL REDUCE, THEN LOSE REDUCTION AFTER TX RELEASED
  - SOME WILL NOT REDUCE AT ALL
  - SURGERY INDICATIONS (C1-2 PSF)
    - FIXED DEFORMITY
    - LOSS OF REDUCTION AFTER TREATMENT
    - ANT DISPLACEMENT > 5mm
    - NEURO SX
    - >3 MONTH DURATION OF SYMPTOMS
KLIPPEL-FEIL SYNDROME

- Congenital fusions of cervical vertebrae
- Clinical triad:
  - Low posterior hairline
  - Short neck
  - Limited neck motion
- Abnormal embryologic development of vertebrae
- Incidence = 0.7%
KLIPPEL-FEIL SYNDROME

• SPRENGEL DEFORMITY (33%)
• CARDIAC ANOMALIES
• RENAL ANOMALIES
  – Get renal U/S
• PULMONARY ANOMALIES
• DEAFNESS
• SCOLIOSIS
• X-RAYS VARIABLE:
  – Simple block vertebrae to bizarre anomalies
KLIPPEL-FEIL SYNDROME

- INSTABILITY COMMON ADJACENT TO FUSED LEVELS
  - Get flex/ext views prior to anesthesia
  - Commonly see C1-2 and C3-4 fusion with instability risk at unfused C2-3
KLIPPEL-FEIL SYNDROME

• NO CLEAR TX GUIDELINES
  - Treat similar to congenital scoliosis
  - High risk of developing instability
    • Avoid contact sports
    • Cervical traction, collars, analgesics for mechanical sx
  - Surgery
    • Fuse for neuro signs due to instability
    • Surgery for cosmesis alone = unwarranted and risky
ATLANTO-OCCIPITAL SYNOSTOSIS

• Anterior arch C1 fused to occiput
  - Posterior arch typically hypoplastic

• C1 height variably decreased
  - Allows odontoid to project into foramen magnum
    • “primary basilar impression
    • Often dysplastic odontoid

• Congenital fusion C2-3 in 70%

• Lower C-spine deformities common
ATLANTO-OCCIPITAL SYNOSTOSIS

• FINDINGS: (LIKE KLIPPEL-FEIL)
  - SHORT BROAD NECK
  - DECREASED NECK ROM
  - LOW HAIR LINE
  - SPRENGEL’S DEFORMITY
  - SHORT STATURE
  - HYPOSPADIAS
  - G-U ANOMALIES
  - EAR DEFORMITY
  - CLEFT PALATE
  - JAW ANOMALIES
ATLANTO-OCCIPITAL SYNOSTOSIS

- **OFTEN ASYMPTOMATIC UNTIL 40’S**
- **X-RAYS DIFFICULT TO EVALUATE**
  - Aim beam 90deg to skull, not C-spine
- **CT USUALLY NECESSARY**
  - “head wag” technique in young children
    - Skull blurs, C1-2 articulation visible
- **50% DEVELOP C1-2 INSTABILITY**
  - ADI CAN BE > 12 mm
- **NEURO SX’S CAN DEVELOP SLOWLY**
  - C1-2 instability progresses with age
ATLANTO-OCCIPITAL SYNOSTOSIS

• Neuro symptoms
  – Due to compression of brainstem or anterior upper cord by posteriorly projecting dens
  – Pyramidal signs most common (spastic, hyperreflex, weak, poor gait)
  – Can also get cranial N. involvement, posterior column involvement from foramen lip
ATLANTO-OCCIPITAL SYNOSTOSIS

• TREATMENT
  - C-COLLABRS, BRACES, TRACTION
  - TRAUMA AVOIDANCE
  - SURGERY
    • HIGH RATE OF PARALYSIS
    • WITH C1 - C2 INSTABILITY: C1-2, OR OCCIPUT - C2 FUSION
      - DECOMPRESSION IF NECESSARY
BASILAR IMPRESSION

- Indentation of skull floor by upper C-spine
- Cephalad tip of dens can protrude into foramen magnum
  - Brainstem compression
  - Vascular compromise
  - CSF flow alterations
- Primary vs. secondary

ADI = 11mm

McGregor's > 6.6mm
BASILAR IMPRESSION

- Primary basilar impression
  - More common
  - Congenital abnormality
  - Often assoc with other vertebral defects
    - Klippel Feil
    - Abnormal odontoid
    - Atlas hypoplasia
    - AO fusion
  - 1% incidence
BASILAR IMPRESSION

- Secondary basilar impression
  - Developmental
  - Less common
  - Due to softening of the occiput
    - PAGETS
    - OSTEOMALACIA
    - RICKETS
    - RENAL OSTEOODYSTROPHY
    - OSTEOGENESIS IMPERFECTA
    - ACHONDROPLASIA, HYPOCHONDROPLASIA
    - JRA
    - ANKYLOSING SPONDYLITIS
    - NEUROFI BROMATOSIS
BASILAR IMPRESSION

• FINDINGS
  - SHORT NECK (78%)
  - FACIAL ASSYMMETRY
  - PAINFUL/DECREASED CERVICAL MOTION (53%)
  - NEUROLOGIC SX’S
    • MOTOR WEAKNESS, LIMB PARES ThESIS AS
    • OFTEN ELICITED WITH MINOR TRAUMA
    • WEAKNESS/ PARAESTHESIA
    • OFTEN ASSOCIATED WITH CHIARI
    • CRANIAL N INVOLVEMENT (V, IX, X, XII)
    • HYDROCEPHALUS FROM CSF BLOCKAGE
BASILAR IMPRESSION

- **RADIOLOGY**
  - **PLAIN FILMS DIFFICULT TO ASSESS**
    - **McGREGORS BEST FOR SCREENING**
      - LANDMARKS EASY TO SEE AT ALL AGES ON LATERAL FILM
      - HARD PALATE → OCCIPUT
    - **McRAE BEST FOR MEASURING CLINICAL SIGNIFICANCE**
      - DEFINES FORAMEN OPENING
      - DENS ABOVE LINE = SX
  - CT - BONY ANATOMY, DEGREE OF INVAGINATION
  - MRI - NEURAL ANATOMY/COMPRESSION

Hard palate
BASILAR IMPRESSION

- **TREATMENT**
  - Difficult, requires multidiscipline approach
    - Ortho, neurosurgery, neuroradiology
  - Treatment is surgical
    - PSF in extension at OC junction
    - Anterior excision of odontoid if can’t be reduced
    - May need suboccipital decompression
FAMILIAL CERVICAL DYSPLASIA

- Atlas deformity
- Epidemiology unknown
- Partial absence of C1
  - Usually posterior elements
- Presentation
  - Torticollis, cervical “clunk”, suboccipital pain, decreased ROM
- Radiographs difficult to read
- Instability = Occiput-C1
  - Get flex/ext MRI
FAMILIAL CERVICAL DYSPLASIA

• TREATMENT
  – SERIAL FOLLOW-UP
    • Q6-12mos
  – SURGERY FOR:
    • PAIN
    • INSTABILITY
    • PROGRESSIVE DEFORMITY
    • NEUROLOGIC SYMPTOMS
    • PSF OCCIPUT TO C2 WITH HALO
HEMI-ATLAS

• 3 TYPES
  – I. ISOLATED HEMI-ATLAS
  – II. HEMI-ATLAS WITH LOWER SPINAL ANOMALY
  – III. HEMI-ATLAS WITH ATLANTO-OCCIPITOL ARTHRODESIS

• OFTEN ASSOC. W:
  – FORAMEN MAGNUM STENOSIS
  – CHIARI
  – VERTEBRAL ARTERY ANOMALY
SANDIFER SYNDROME

• GERD with torticollis
• Neck tilt likely attempt to ease reflux discomfort
• Can be diagnosis of exclusion
  - No tight SCM
  - X-rays of c-spine normal
  - Upper GI shows hiatal hernia, GERD
• Treating GERD usually resolves torticollis