Chiari e instabilità cranio-cervicale: aspetti radiologici

Dott. Andrea Garnero

AISMAC
ASSOCIAZIONE ITALIANA SIRINGOMIELIA E ARNOLD CHIARI

SALUS SCIENTIA
AZIENDA OSPEDALIERO - UNIVERSITARIA
Città della Salute e della Scienza di Torino

Acquired disorders

- Inflammatory (Grisel Sd, Juvenile idiophatic arthritis)
- Traumatic
- Tumoral (intramedullary, extramedullary intradural, extradural)
- Iatrogenic (post-laminectomy)

Craniocervical junction instability

Congenital/Developmental disorders

- CCJ malformations
- Down syndrome, Mucopolysaccharidoses, Skeletal dysplasias, Connective tissue disorders, etc.

Imaging techniques

- Plain radiographs (static - dynamic)

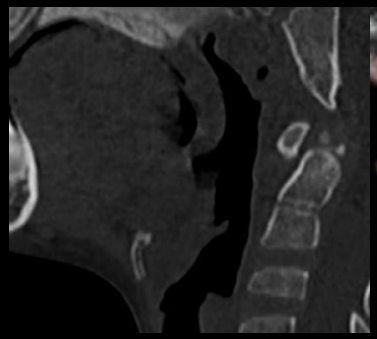
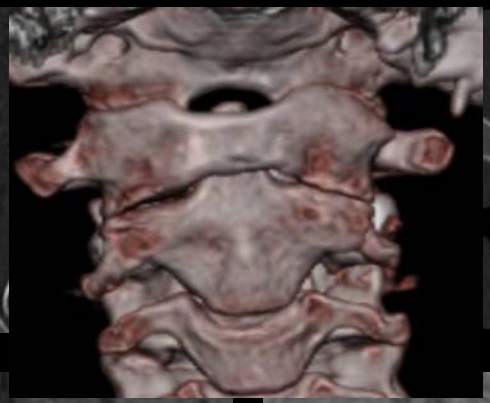
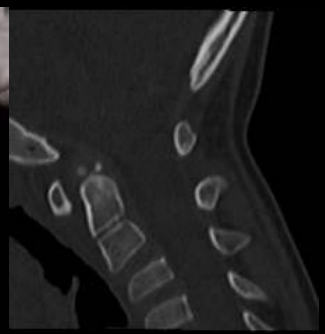
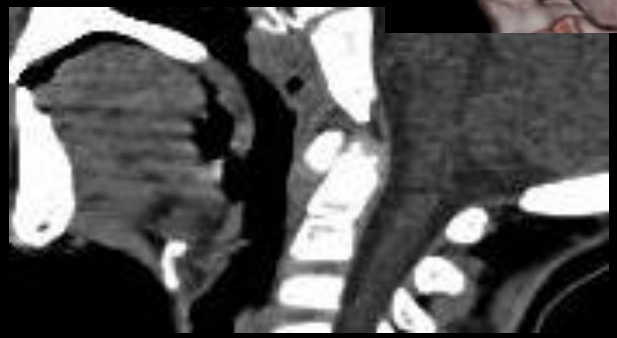
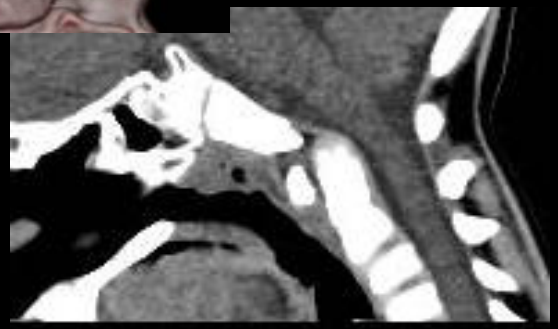
evaluate specific distances between structures of the CCJ



- CT (static - dynamic)

optimally defines the osseous structures

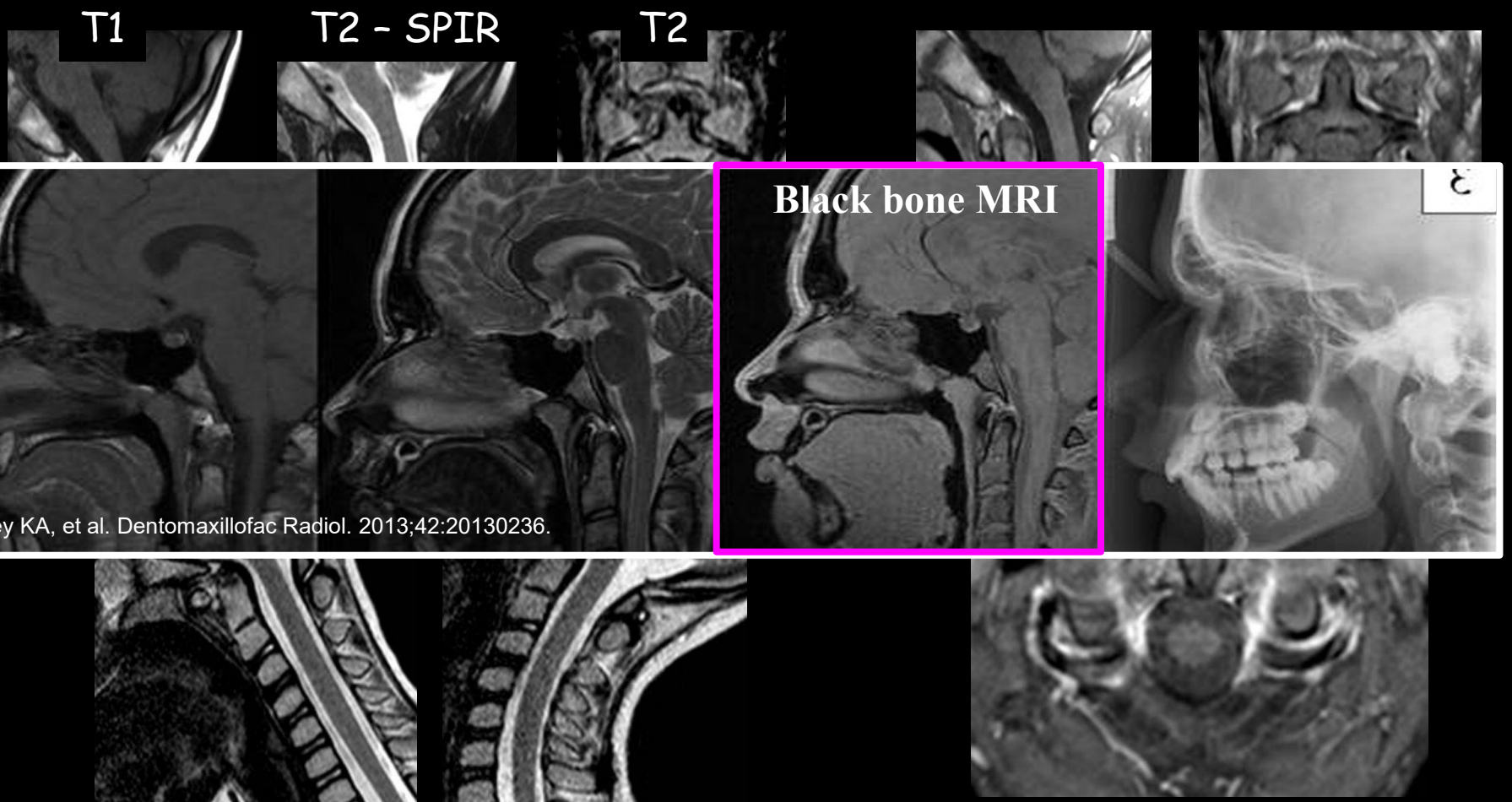
RADIATION EXPOSURE!!!

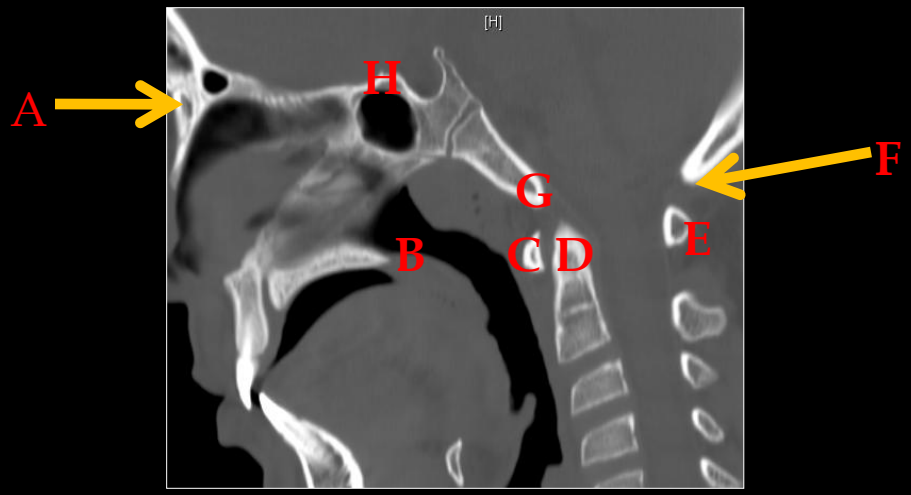
Imaging techniques

- MRI (static - dynamic)

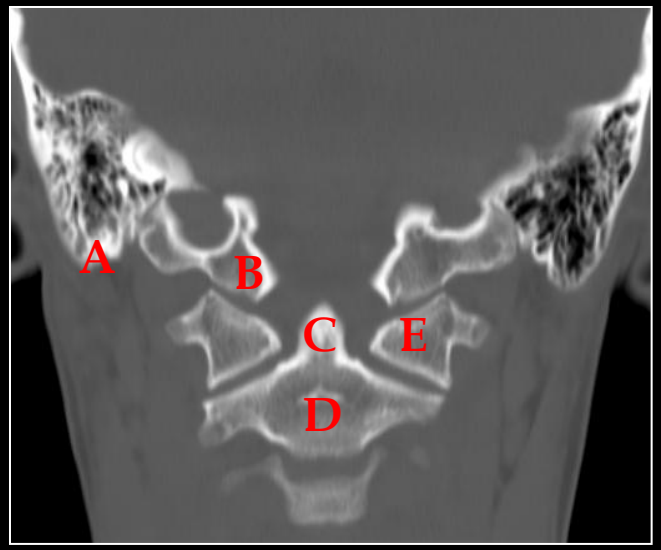
ideal for evaluation of soft tissues, neural structures, and ligaments / bone edema



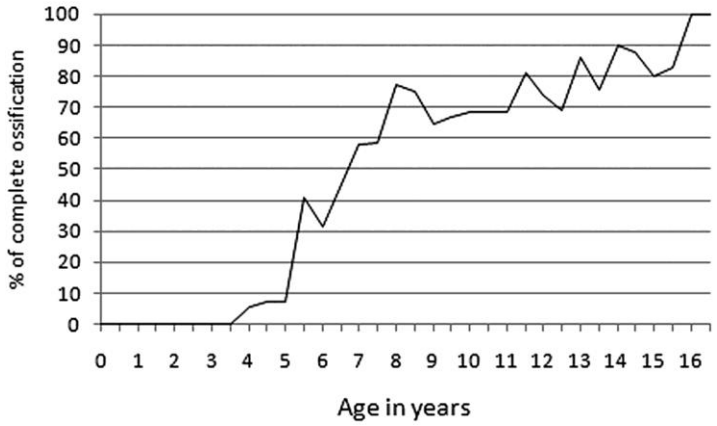
Craniometry - Landmarks

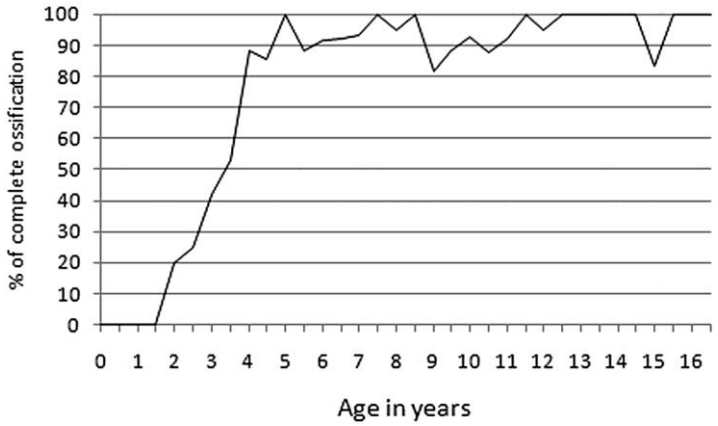


A- nasion
B- posterior pole of hard palate
C- anterior arch C1
D- odontoid process
E- posterior arch C1
F- opisthion
G- basion
H- tuberculum sellae



A- tip of mastoid process
B-occipital condyle
C-odontoid process
D-axis body
E-lateral mass of atlas





AJNR
AMERICAN JOURNAL OF NEURORADIOLOGY
Username Password GO
Advanced Search
Institution: ARS Infor_Biomed Liguria

AJNR Am J Neuroradiol 33:1882–87 | Nov 2012 | www.ajnr.org

ORIGINAL RESEARCH

Normal Ossification Patterns of Atlas and Axis: A CT Study

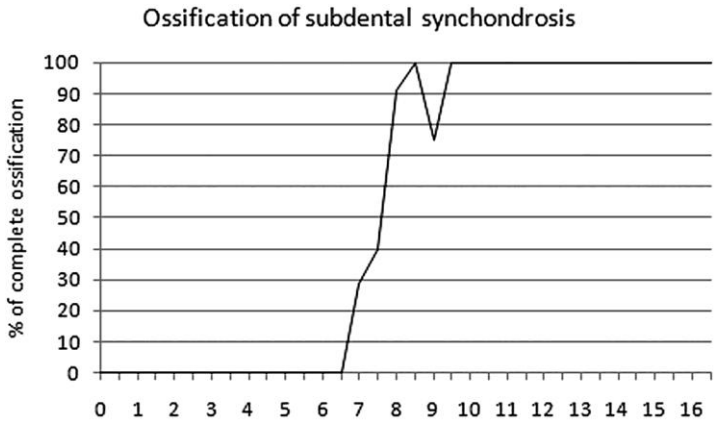
G.M. Karwacki
J.F. Schneider

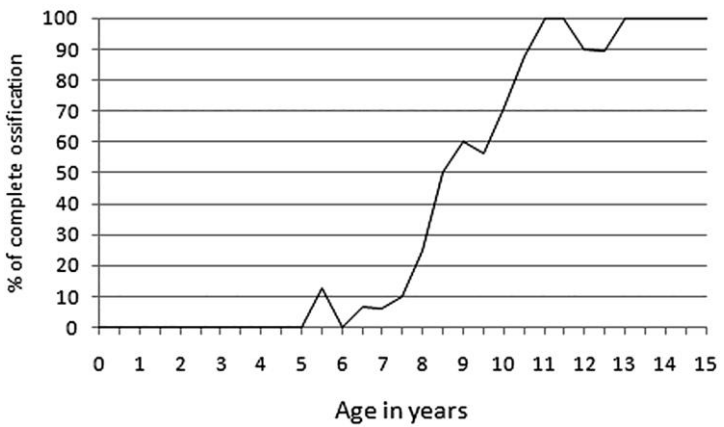
BACKGROUND AND PURPOSE: Development of the CVJ is a complex process rarely analyzed by CT. Cartilaginous remnants within the atlas and axis have been shown to variably persist throughout childhood and may be mistaken for fractures. The purpose of this study was to better estimate the fusion timeline of the synchondroses at the CVJ.

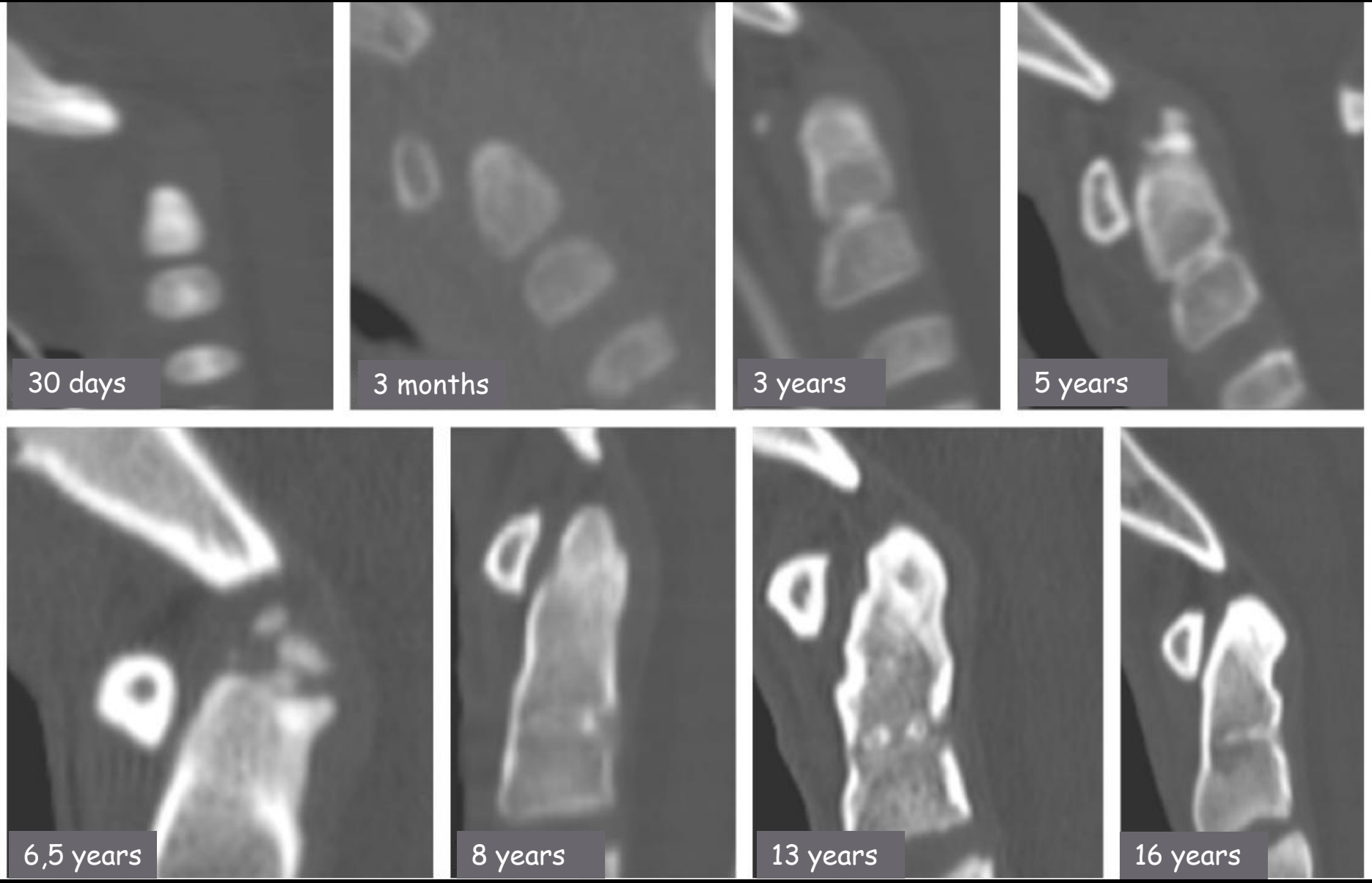
MATERIALS AND METHODS: We retrospectively reviewed singular CT scans in 550 children without known skeletal dysplasia or maturation delay (from neonate to 17 years) and analyzed the ossification of atlas and axis. Normal closing age for synchondroses was defined as soon as a complete ossification rate of 80% or more per age category was achieved.

RESULTS: No separate OC was observed in 38 of 230 incompletely ossified AAAs, whereas single, bipartite, or multiple OCs were observed in, respectively, 196, 86, and 16 of 298 AAAs in which OCs could be identified. Synchondroses at the AAA closed after 12 years (range 4.5–17 years). Posterior midline synchondrosis of the atlas closed after 4 years (range 2–13 years). The axis ossified from 6 OCs and 4 synchondroses. Subdental and neurocentral synchondroses closed simultaneously after 9 years (range 7–9.5 years). Apicodental synchondrosis and chondrum terminale were completely ossified after 10.5 years (range 5.5–13.5 years).

CONCLUSIONS: Analysis of the CVJ using triplanar CT reconstructions allows secure identification of characteristic developmental features of the atlas and axis, and helps to separate normal variants from true osseous lesions.







G.M. Karwacki
J.F. Schneider

Normal Ossification Patterns of Atlas and Axis: A CT Study

AMERICAN JOURNAL OF NEURORADIOLOGY

CCJ instability

Instability occurs each time the anatomical relationships between the articular components are altered or have the propensity of becoming abnormal under the influence of certain solicitations

→ risk of neurological involvement

Broken red circle

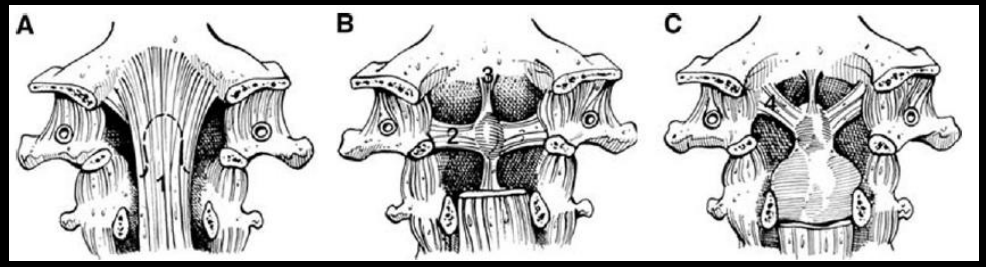
- Atlanto-occipital dislocation (C0-C1 instability)
- Atlanto-axial subluxation (C1-C2 instability)
- Sub-axial subluxation

Atlanto-occipital dislocation

Malalignment betweeen the cranium and the spine (C0-C1)

Rare (post-traumatic/congenital disorders)

Rupture of ligaments/ ligamentous laxity



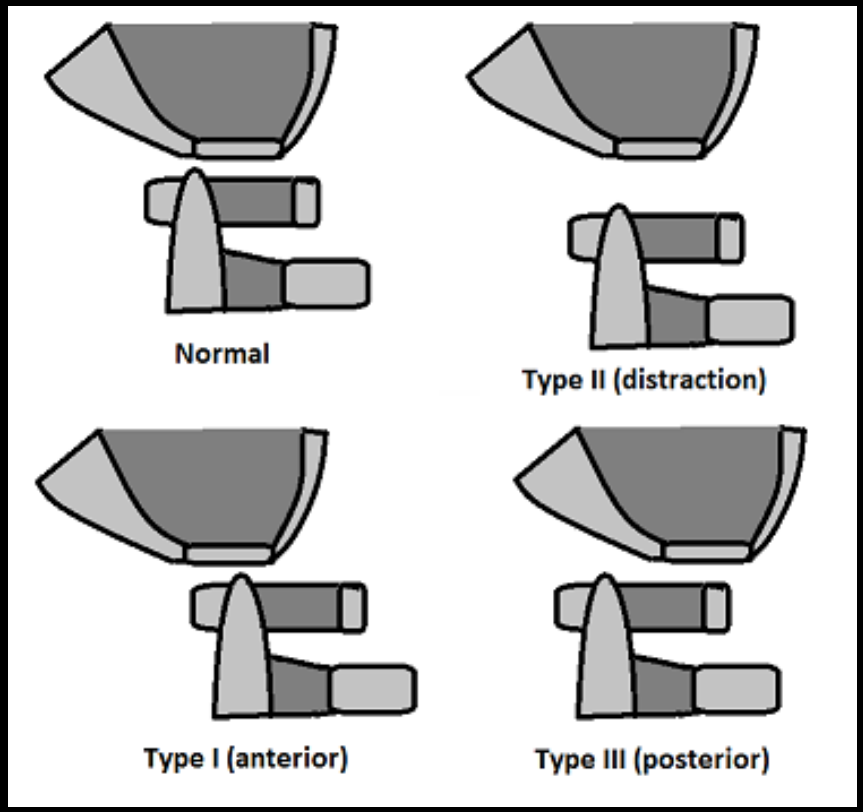
J Child Orthop (2008) 2:71–84
DOI 10.1007/s11832-008-0092-2

CURRENT CONCEPT REVIEW

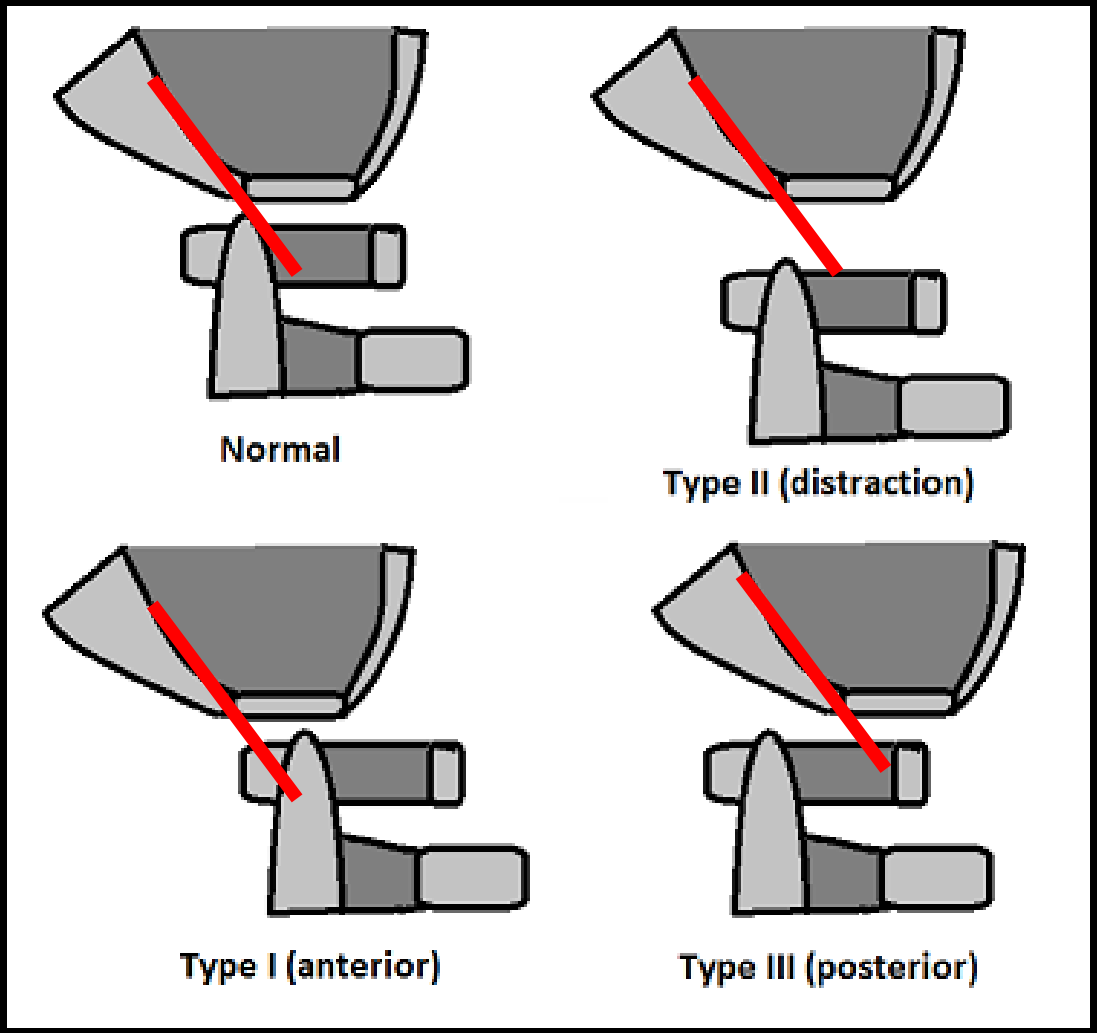
Pediatric cervical spine instability

Ismat Ghanem · Samer El Hage · Rami Rachkidi · Khalil Kharrat · Fernand Dagher · Gabi Kreichati

Traynelis classification



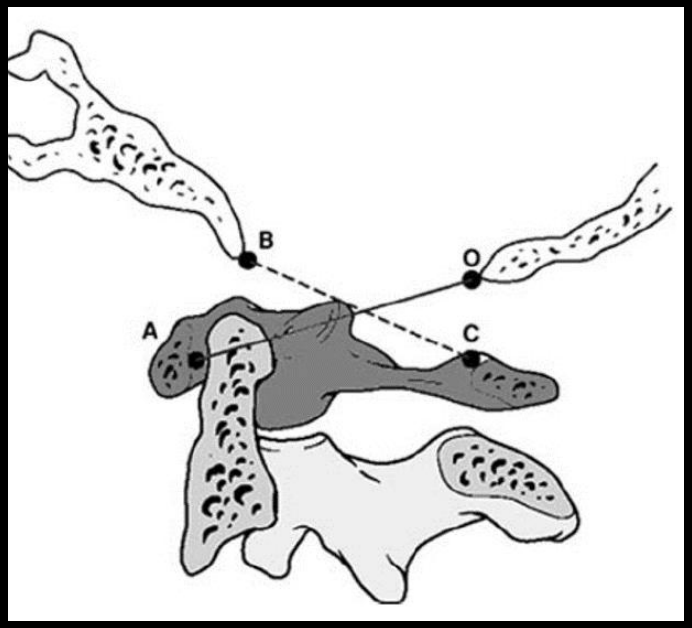
Atlanto-occipital dislocation

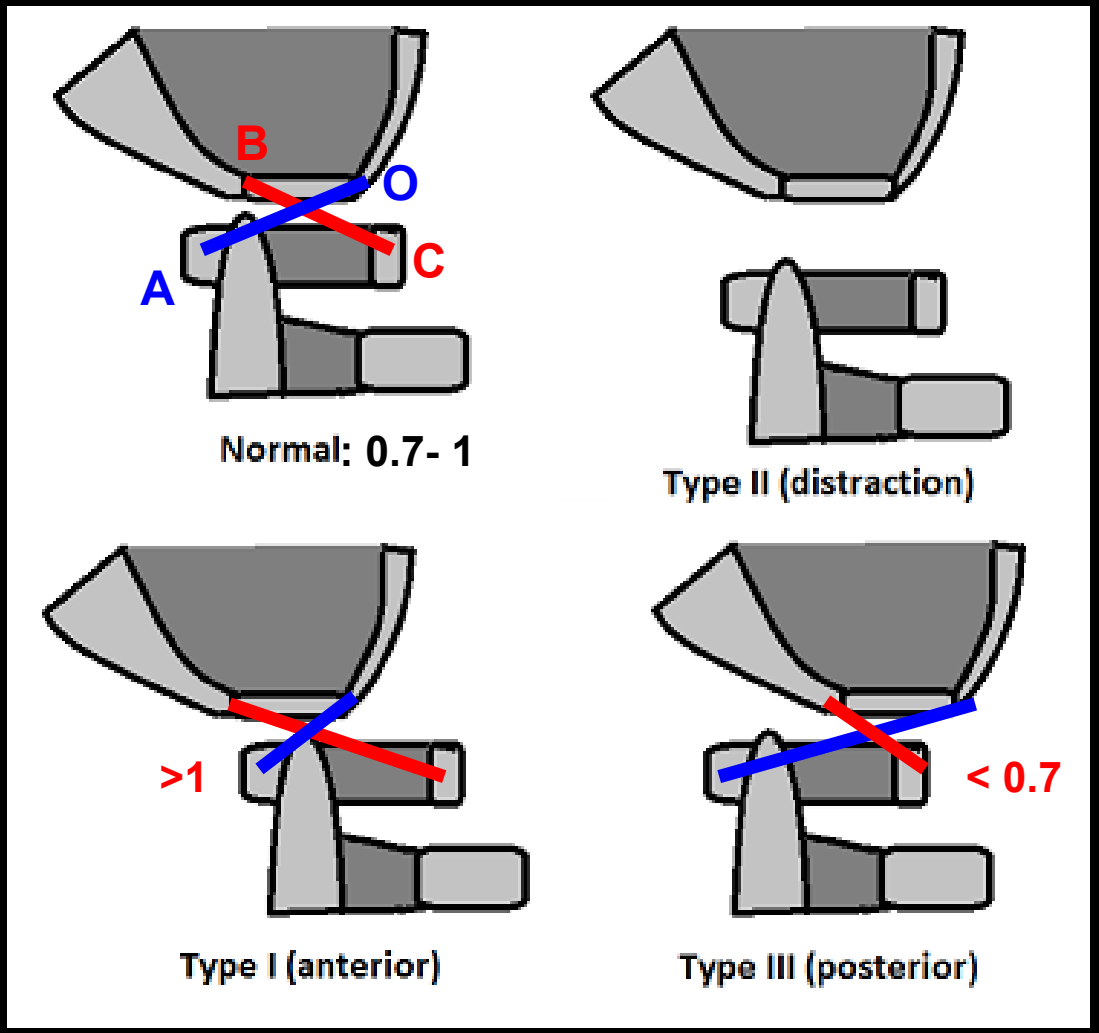


Wackenheim's clivus baseline: should fall tangent to, or intersect, the posterior one third of the odontoid

Atlanto-occipital dislocation

used to assess the position of the skull base relative to the atlas

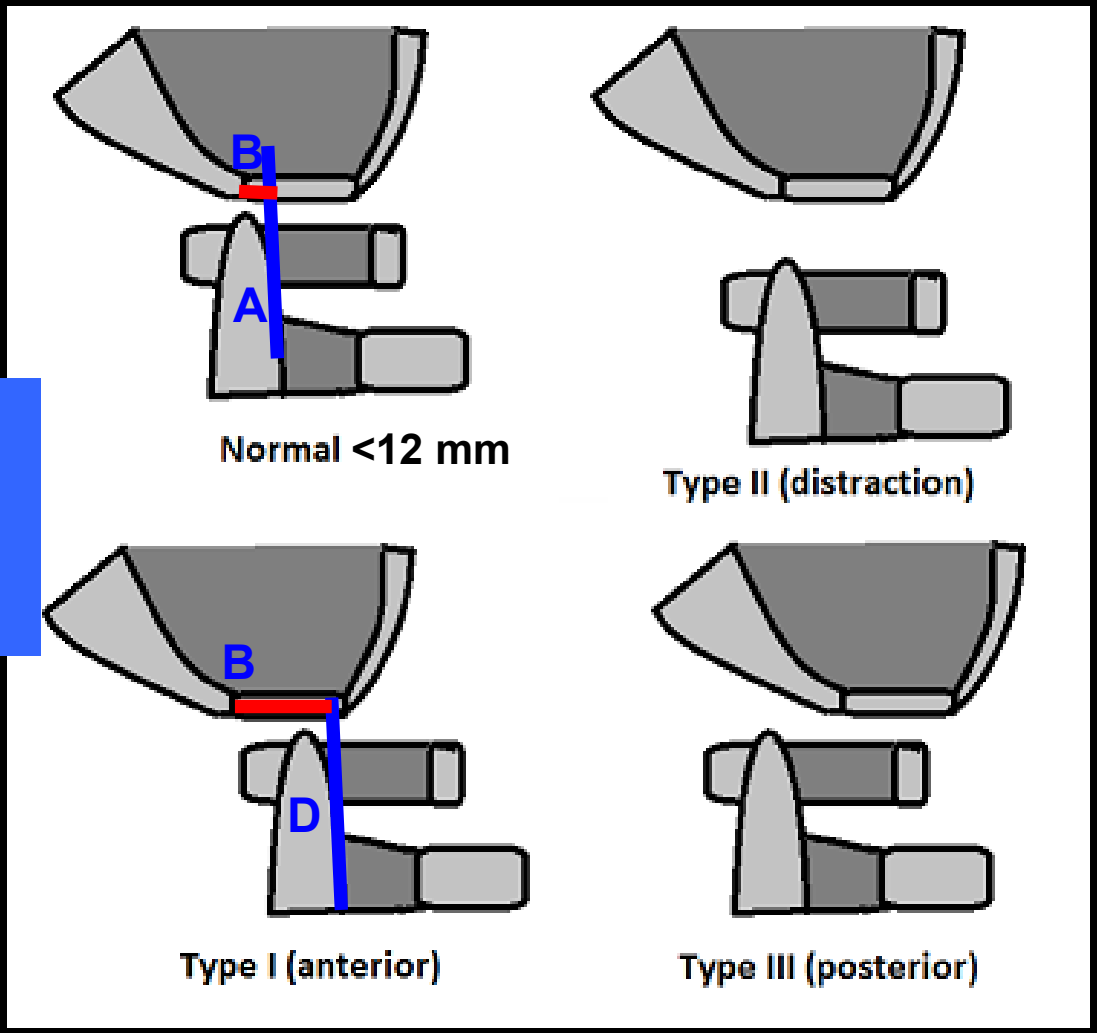




Powers ratio: the ratio BC/OA should be between 0,7 and 1
Sensitive for type I and III injury

Atlanto-occipital dislocation

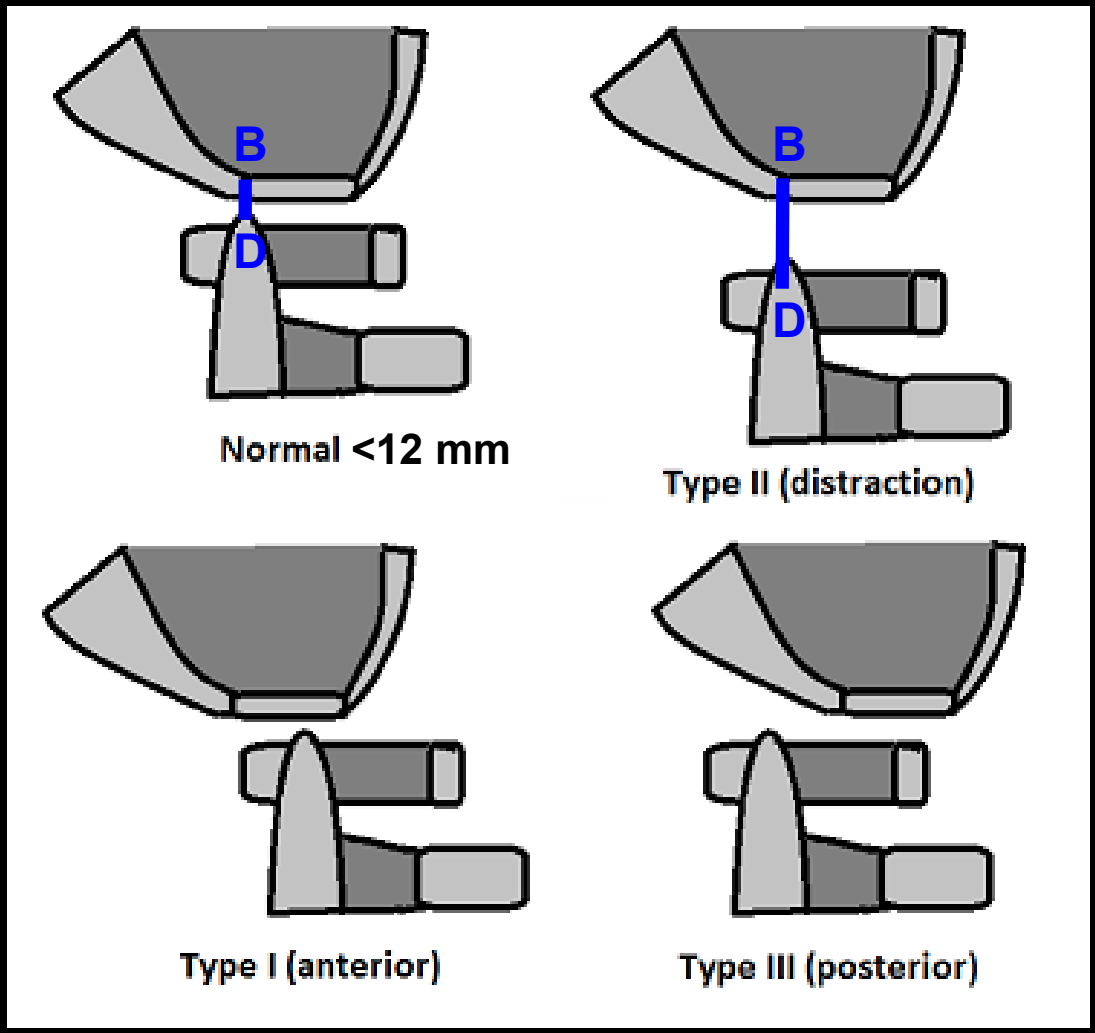
Horizontal Harris Measurement (HHM)



Basion axis interval: distance between the basion and the superior extension of the posterior cortical margin of the body of the axis should be less than 12 mm

Sensitive for type I injury

Atlanto-occipital dislocation



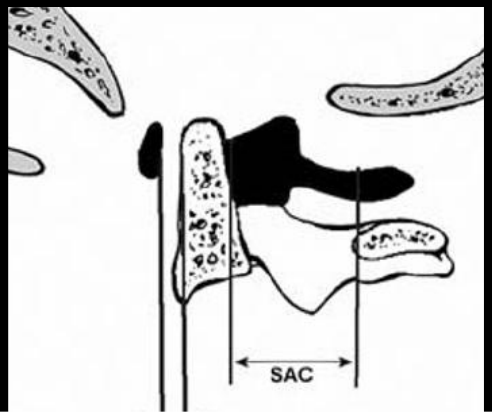
Basion dens interval: distance between the basion and the tip of the odontoid should be less than 12 mm in children

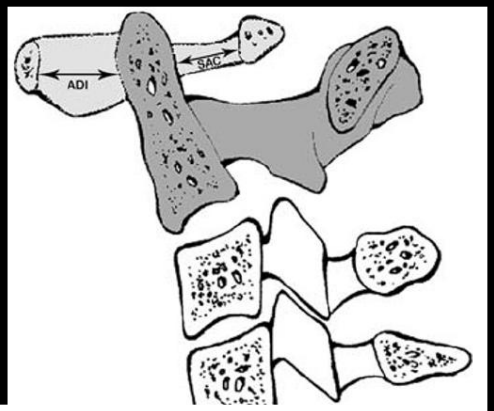
Sensitive for type II injury

Atlanto-axial dislocation (C1-C2 instability)

Anterior atlantodens interval (AADI) and space available for the cord (SAC - PADI)

AADI : < 5 mm in children (<8 years old)





Sagittal Diameter of the Cervical Spinal Canal in Children

A SAC of less than 13 mm is significant

Diameter	Sex	Age	Mean	S.D.	90% Tolerance Range Lower Limit (P_{05})	90% Tolerance Range Upper Limit (P_{95})
C-1	Boys (N = 27)	3	19.5	1.7	15.2	23.8
		8	20.0	1.7	15.7	24.3
		13	20.7	1.7	16.4	25.0
		18	21.3	1.7	17.0	25.6
	Girls (N = 21)	3	16.8	1.5	12.9	20.7
		8	17.8	1.5	13.9	21.7
		13	18.8	1.5	14.9	22.7
		18	19.7	1.5	15.8	23.6

Radiology. 1962;79:97-108

An AADI with **> 2 mm difference between flexion and extension: instability**

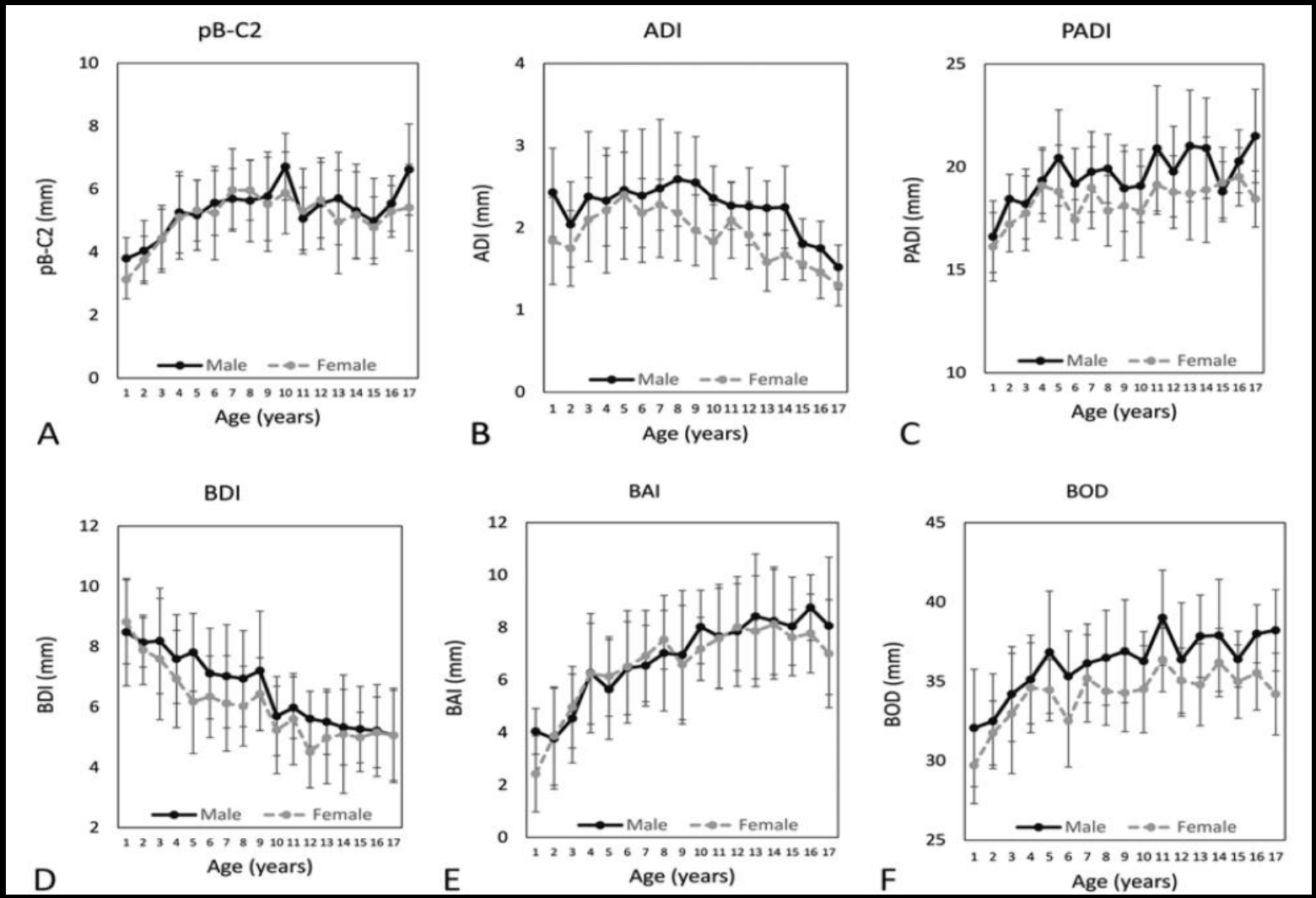
In younger children plain radiographic measurements of ADI and SAC are not very reliable for assessment of instability or stenosis, and cord compression cannot be detected: MRI

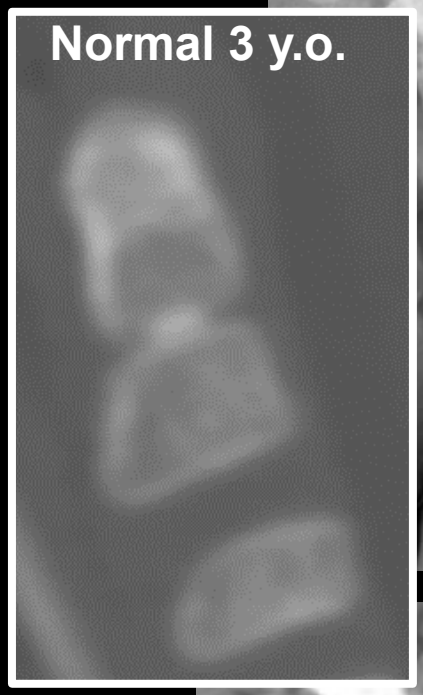
Morphometric changes at the craniocervical junction during childhood

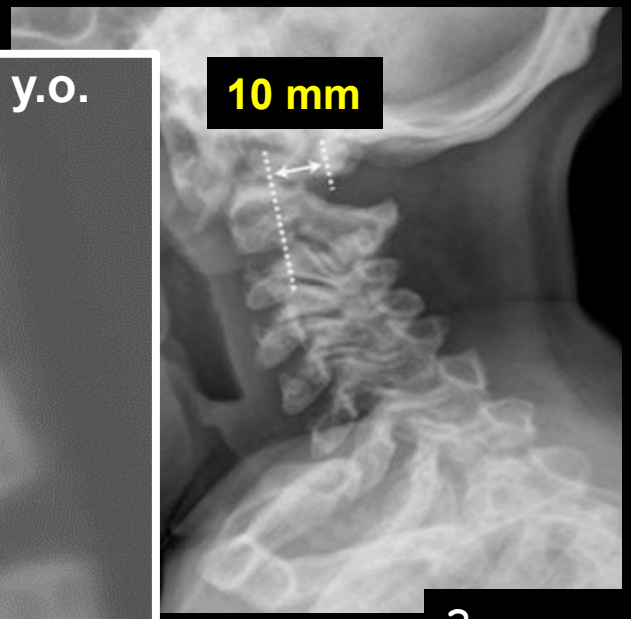
J Neurosurg Pediatr 24:227–235, 2019

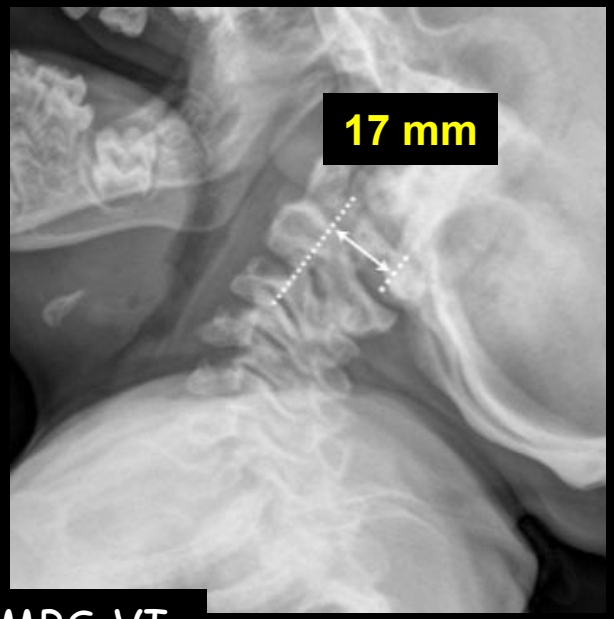
*Jayapalli Rajiv Bapuraj, MD,[1] Amy K. Bruzek, MD,[2] Jamaal K. Tarpeh, BS,[2] Lindsey Pelissier, MD,[2] Hugh J. L. Garton, MD, MHSc,[2] Richard C. E. Anderson, MD,[3] Bin Nan, PhD,[4] Tianwen Ma, MS,[5] and Cormac O. Maher, MD[2]

1458 eligible patients were identified from children between 1 and 18 years of age who underwent cervical spine CT scanning

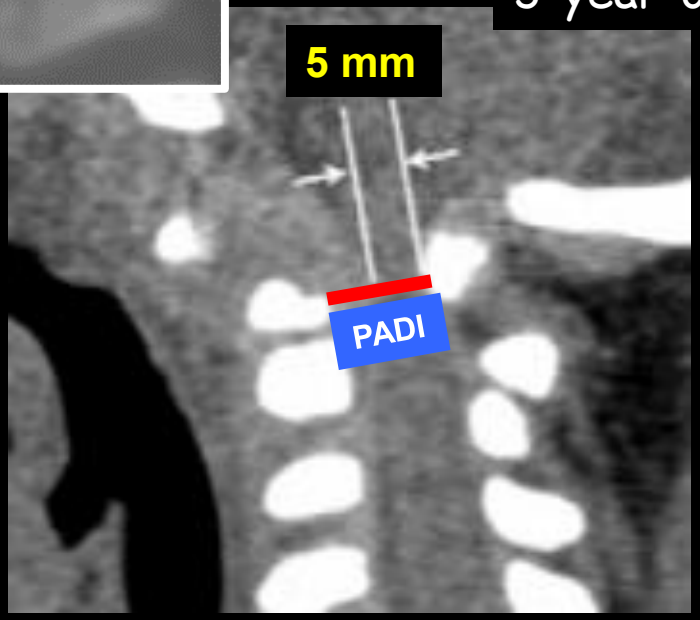








3-year-old MPS VI



G.A. Solanki et al. / Molecular Genetics and Metabolism 118 (2016) 310–318

Congenital/Developmental disorders
CCJ malformations

Dachling Pang · Dominic N. P. Thompson
Embryology and bony malformations of the craniovertebral junction
Childs Nerv Syst (2011) 27:523–564

Malformations of central pillar
Dens, C2 body, and basiocciput

Disturbance of axial component of occipital sclerotome, proatlas and C_1 resegmented sclerotome

Odontoid dysgeneses
- Aplasia/hypoplasia of odontoid components
- Disturbance of odontoid synchondroses (IBZ)
  - Os odontoideum
  - Ossiculum terminale persistens
- Abnormal resegmentation of proatlas centrum
  - Os avis
- Failed midline integration of basal dental segment
  - Bifid dens

Basioccipital dysgeneses
- Failed midline integration of basioccipital primordium
  - Bifid clivus
- Basioccipital dysplasia
  - Basilar impression
  - Platybasia
  - Retroflexed dens
  - Basilar invagination
  - Basilar kyphosis

Malformations of surrounding rings
Foramen magnum and atlantal rings

Disturbance of lateral component and hypochordal bows of proatlas and C_1 resegmented sclerotome

Proatlas anomalies
- Hyperplasia of hypochordal bow of proatlas
  - Third occipital condyle
  - Pre-basioccipital arch
- Hyperplasia of exoccipital sclerotome
  - Hypertrophic occipital condyle
- Non-resegmentation of proatlas (anterior homeotic transformation)
  - Atlas assimilation
- Posterior homeotic transformation

C_1 sclerotome anomalies
- Aplasia of hypochordal bow of C_1
  - Aplasia and hypoplasia of anterior C_1 arch
- Aplasia/hypoplasia of lateral sclerotome
  - Posterior C_1 arch agenesis
- Combined hypochordal bow and lateral sclerotome dysplasia
  - Aplasia of lateral mass and anterior C_1 arch
  - Combined anterior and posterior C_1 arch defects
  - Bifid anterior and posterior C_1 arch

Associated condition

Chiari I

instability

neural compression

Chamberlain's line:

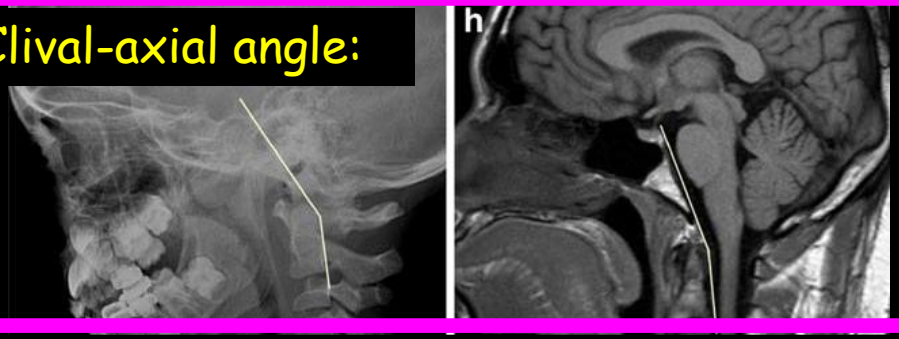
posterior margin of hard palate to opisthion

Smoker W, Childs Nerv Syst (2008)

Tip of the odontoid should be no more than 5 mm above this line

Clivus-canal angle or Clival-axial angle:

Angle formed at junction of Wackenheim's line and posterior vertebral body line



Range= 150 to 180°

Abnormal if:
<135° (Henderson et al.)
<125° (Brockmayer et al.)

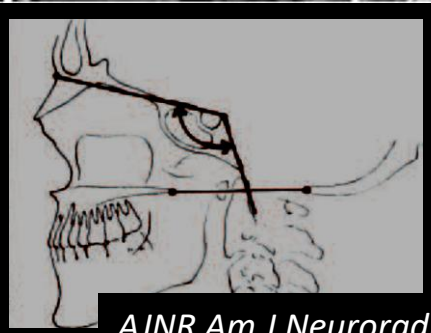
Basal angle:

Angle subtended by the junction of the nasion-tuberculum and tuberculum-basion tangents

Should be less than 140°

McGregor

Variation of 2-3° less



«modified angle»

Normal range 104°-124° for children

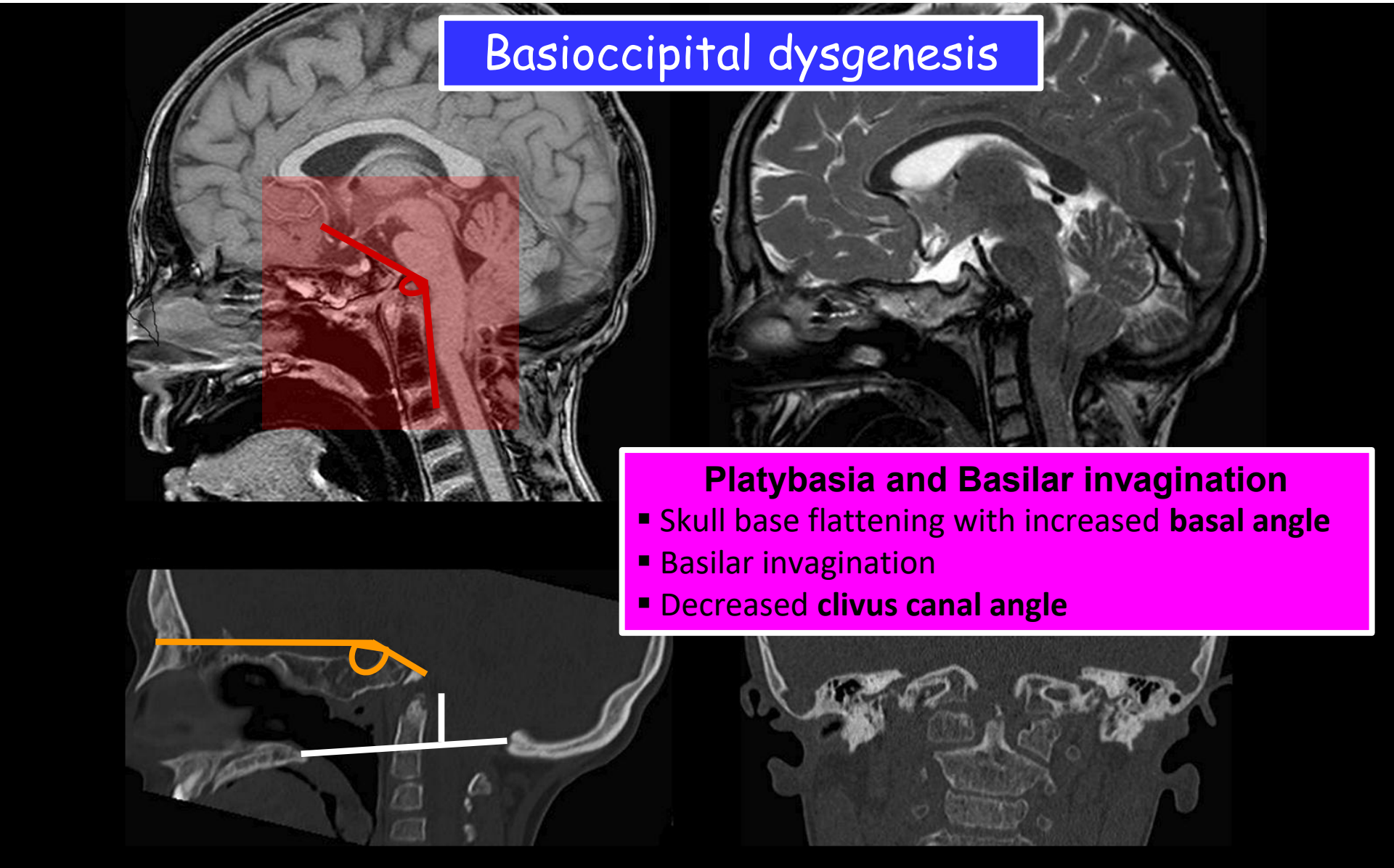
AJNR Am J Neuroradiol 26:89–92, 2005

Tonsillar herniation spectrum: more than just Chiari I. Update and controversies on classification and management

Neurosurgical Review
https://doi.org/10.1007/s10143-019-01198-x

REVIEW 2019

Pietro Fiaschi[1,2] · Giovanni Morana[3] · Pasquale Anania[1,4] · Andrea Rossi[3] · Alessandro Consales[5] · Gianluca Piatelli[5] · Armando Cama[5] · Marco Pavanello[5]



Neurosurg Clin N Am 26 (2015) 561–569

Craniovertebral Junction Instability in the Setting of Chiari I Malformation

Hannah E. Goldstein, MD*, Richard C.E. Anderson, MD

Most children with symptomatic CMI, even those presenting with signs of brainstem compression, do not have CVJ instability and will improve with a standard posterior fossa decompression alone.

Neurol Sci (2011) 32 (Suppl 3):S345–S347
DOI 10.1007/s10072-011-0690-5

ASSOCIATED MALFORMATIONS

The complex Chiari: issues and management strategies

Douglas L. Brockmeyer

cerebellar tonsil herniation combined with one or more of the following radiographic findings: Chiari 1.5 malformation, medullary kink, retroflexed odontoid, abnormal clival cervical angle, occipitalization of the atlas, basilar invagination, syringomyelia

Neurosurg Clin N Am 26 (2015) 561–569

Craniovertebral Junction Instability in the Setting of Chiari I Malformation

Hannah E. Goldstein, MD*, Richard C.E. Anderson, MD

- Defining CVJ instability in the setting of Chiari I is often challenging
- There is limited consensus among neurosurgeons about the exact clinical and radiographic criteria that should be used when evaluating patients with Chiari I to determine which patients should undergo CVJ stabilization

The diagnosis of CVJ instability should be based both on clinical and radiographic findings

Clinical findings associated with CVJ instability in the setting of Chiari I are often not distinct from the symptoms attributable to the Chiari malformation

Clinical findings suggestive of craniovertebral junction instability in the setting of Chiari I malformation

- Occipital headache
- Neck pain
- Myelopathy
- Hyperreflexia
- Central cord syndrome
- Posterior column dysfunction
- Downbeat nystagmus
- Decreased gag reflex
- Dysphagia

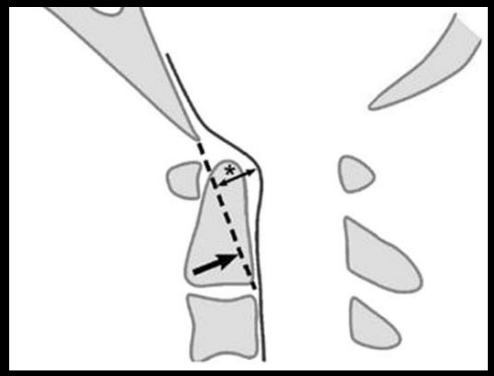
Radiographic findings suggestive of CVJ instability

Neurosurg Clin N Am 26 (2015) 561–569

Craniovertebral Junction Instability in the Setting of Chiari I Malformation

- Basilar Invagination
- Ventral brainstem compression
- Craniocervical Angulation

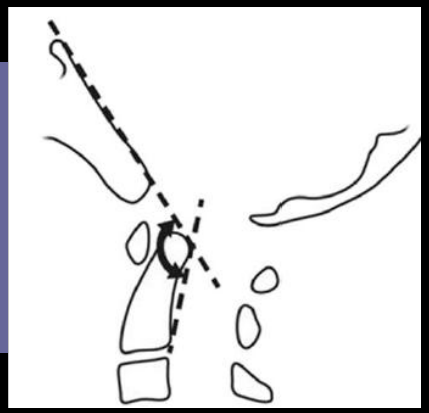
Grabb-Oakes measurement



pB-C2 distance: the distance extending perpendicularly from the B-C2 line through the odontoid tip to the ventral dura

Grabb and colleagues found that if > 9 mm is associated with symptomatic ventral brainstem compression and risk of CVJ instability

Clivus-canal angle less than 125 has been shown to be a significant predictive factor for requiring CVJ fusion



- Chiari 1.5
- Genetic Syndromes (connective tissue disorders)

However…

There is no single radiographic finding that is in itself diagnostic of CVJ inst.

Child's Nervous System
https://doi.org/10.1007/s00381-019-04214-z
2019
SPECIAL ANNUAL ISSUE

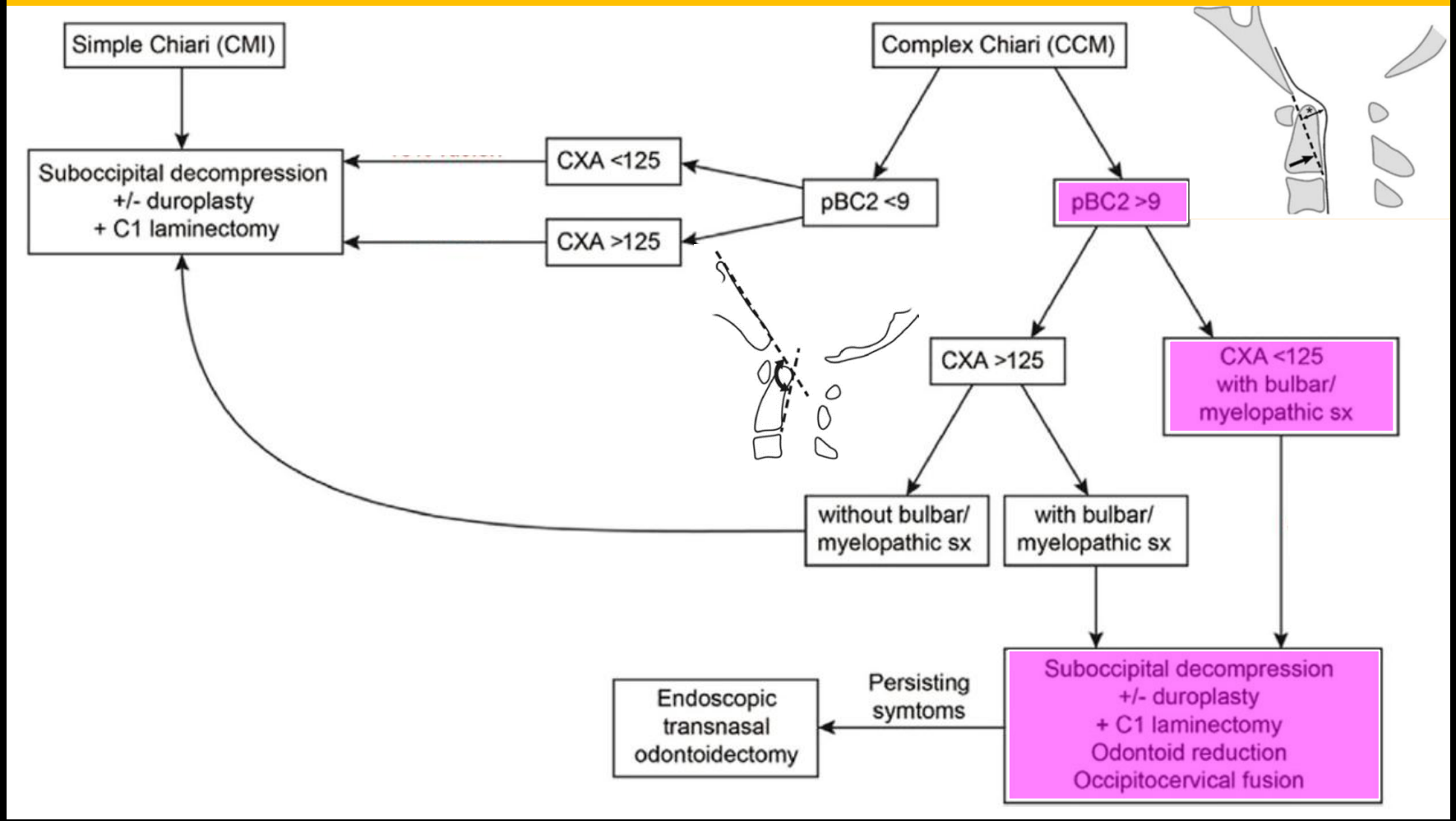
Complex Chiari malformation: using craniovertebral junction metrics to guide treatment

Winson S.C. Ho [1] · Douglas L. Brockmeyer [1]

Radiographic parameters

- pB-C2 distance
- Clival–axial angle (CXA), also known as clivus-canal angle

Algorithmic strategy to identify patients at risk of needing OC fusion



Neurosurgical Review (2019) 42:915–936
https://doi.org/10.1007/s10143-018-01070-4

ORIGINAL ARTICLE

CrossMark

Cervical medullary syndrome secondary to craniocervical instability and ventral brainstem compression in hereditary hypermobility connective tissue disorders: 5-year follow-up after craniocervical reduction, fusion, and stabilization

Fraser C. Henderson Sr[1,2] · C. A. Francomano[1] · M. Koby[1] · K. Tuchman[2] · J. Adcock[3] · S. Patel[4]

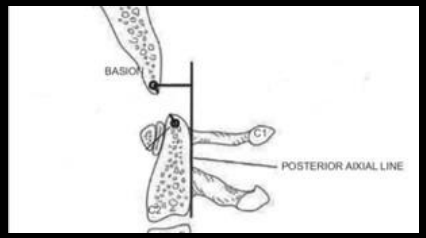
20 consecutive patients with hereditary connective tissue disorders (Ehlers-Danlos syndrome)

Three radiologic metrics used in this study:

- Clival-axial angle (CXA): **< 135°** considered potentially pathological

- pBC2 distance (Grabb-Oakes measurement): > 9 mm

- Basion axis interval: > 12 mm (Horizontal Harris Measurement)



(a change in the basion axis interval of 2 mm or more, as measured in flexion and extension images, represents pathological translation between the basion and odontoid)

Neurological Sciences
https://doi.org/10.1007/s10072-021-05347-3

ORIGINAL ARTICLE

Diagnosis and treatment of Chiari malformation and syringomyelia in adults: international consensus document

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Neurosurgical Review (2019) 42:915–936
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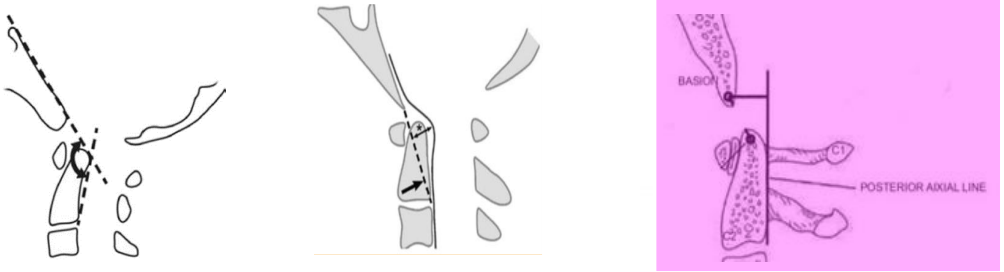

"CM1.5" or "complex Chiari" implies the occurrence of tonsil prolapse and brainstem kinking in the setting of a cranio-vertebral junction (CVJ) malformation (i.e., Klippel-Feil anomaly, atlanto-occipital fusion, basilar invagination, retroversion of the odontoid process) and requires a specific surgical approach.

C3 **CM-1.5**: cerebellar tonsils and brainstem herniation (kinking) below the McRae line; defined "complex" if also skeletal deformities of craniovertebral junction and cervical spine (Klippel-Feil anomaly, atlanto-occipital fusion, basilar invagination, retroversion of the odontoid process) are present.

Table 6 Specific conditions related to CVJ malformations: surgical options

4	Cranio-vertebral junction (CVJ) instability is a mobile dislocation between C0, C1, and C2 (according to quantitative neuro-radiological criteria*) leading to neuro axial compression, neurological deficits, progressive deformity, or structural pain.	Agreement: 94.1% Grade D
5	The standard diagnostic workup for CVJ instability in CM should include (other than MRI) dynamic X-rays + dynamic CT scan with 3D reconstructions	Agreement: 94.1% Grade D

*The clivo-axial angle (angle between clivus line and the posterior axial line, equal or less than 135 degrees), the Harris measurement (distance from the basion to the posterior axial line equal or greater than 9 mm), and the Grabb-Oakes method (draw a line from basion to the posterior inferior edge of the C2 vertebra greater than 9 mm)

(> 12 mm)

Grazie per l’attenzione

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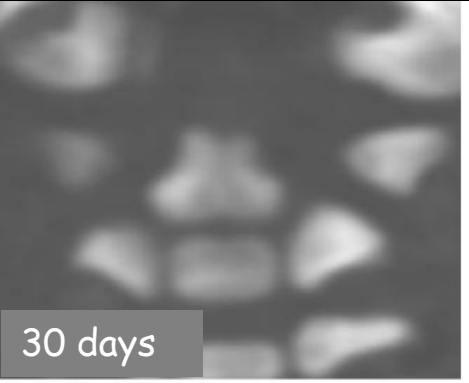
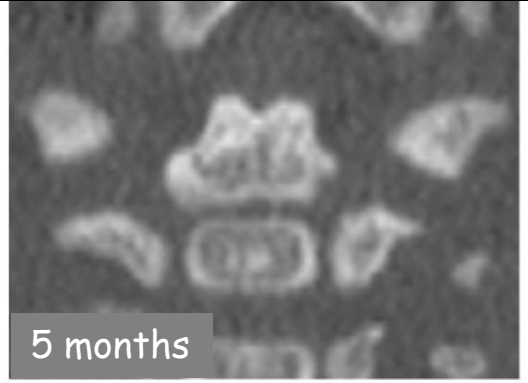
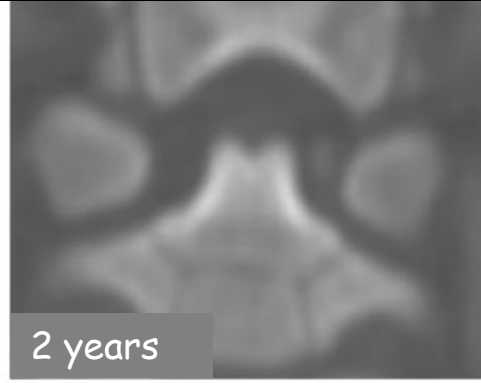
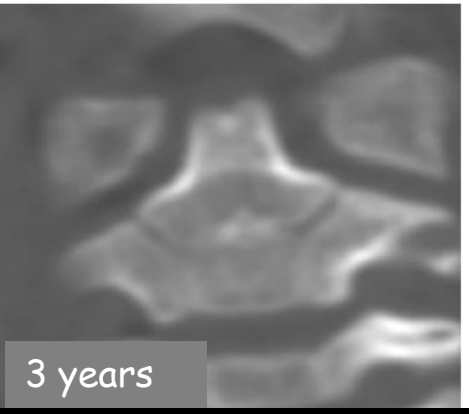
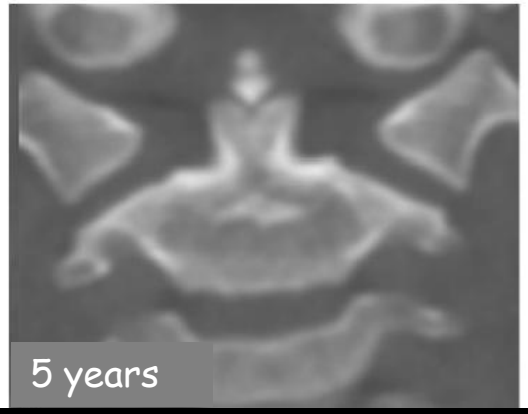
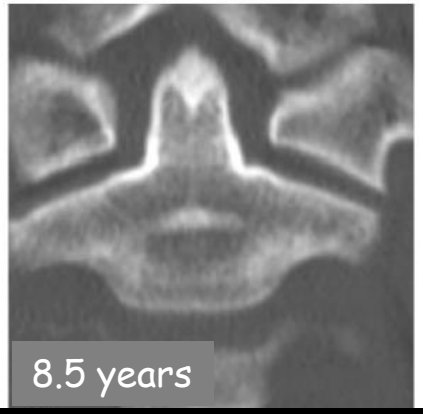




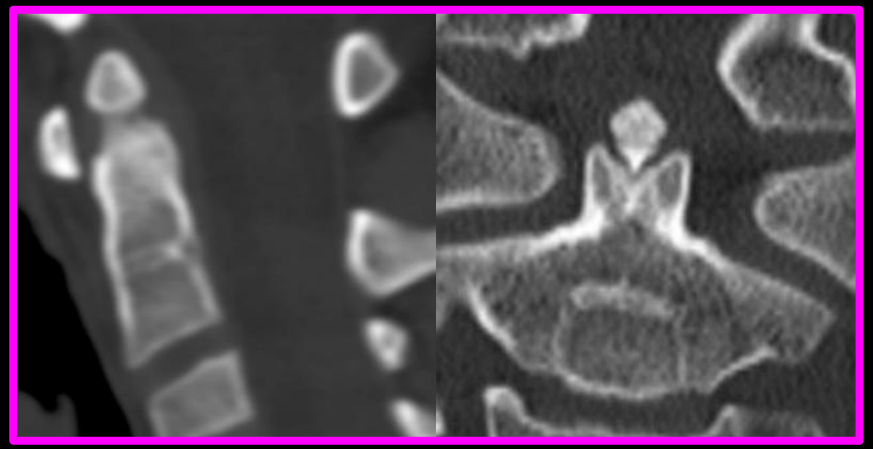








Os odontoideum: separate ossicle superior to the dens



Persistent ossiculum terminale (16 y.o.)