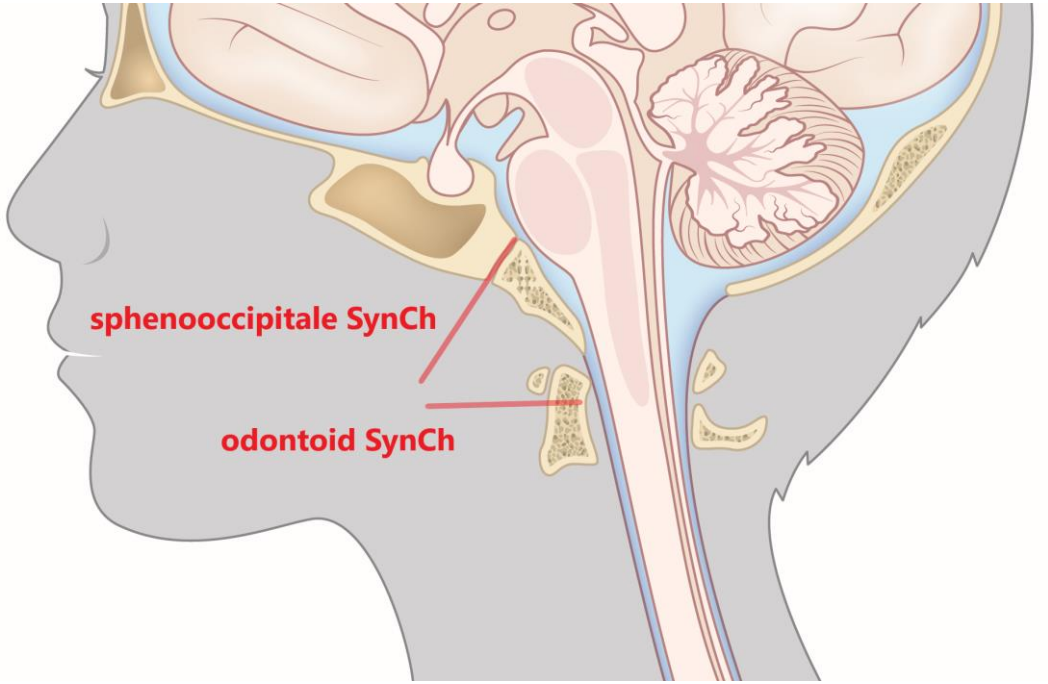
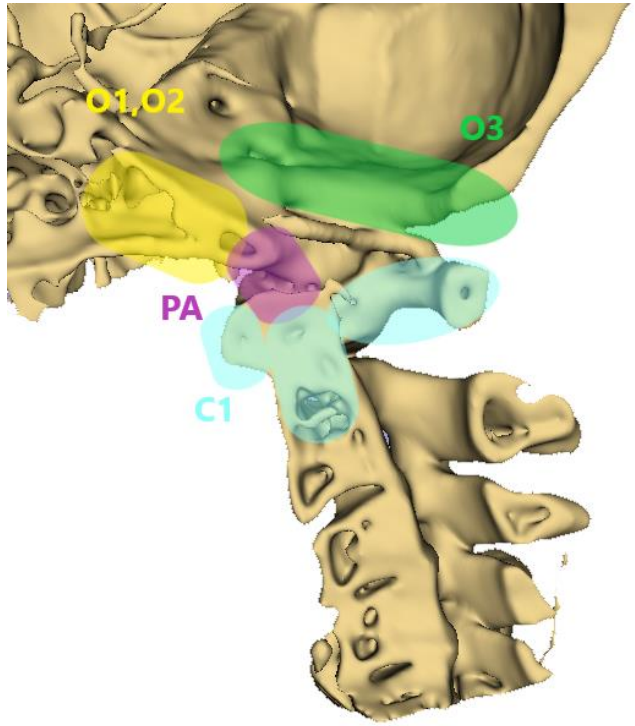
Pathologien des craniocervicalen Überganges bei Kindern

A.Spiessberger

«Craniocervicaler Übergang» - Definition

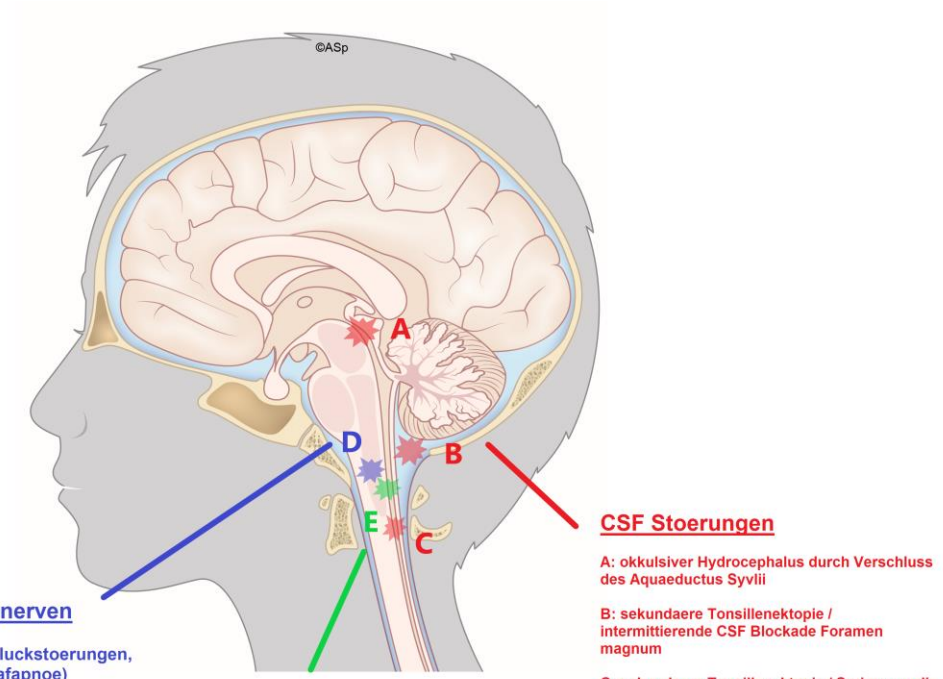




Sklerotome Occ1-3, Proatlas, Cer1

Mögliche Symptome

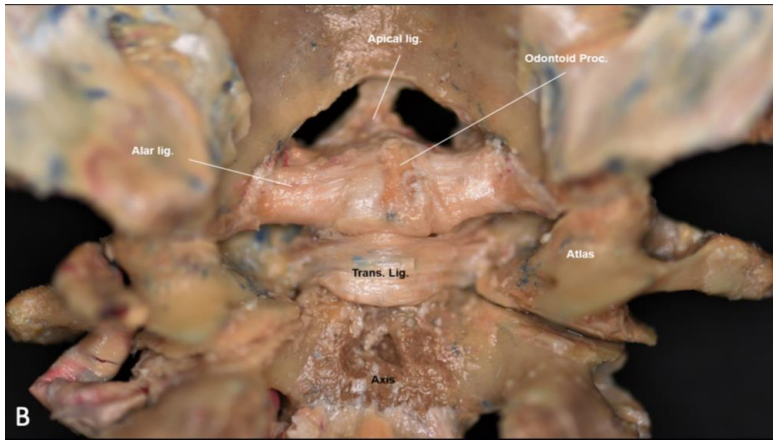
Symptome



sekundäre Veränderungen

- Sekundäre Tonsillenektopie (Syringomyelie, Syringobulbie)
- Okkulsiver Hydrocephalus (Foramen Magendi)

Pathophysiologie



Insuffizienz TAL-Komplex

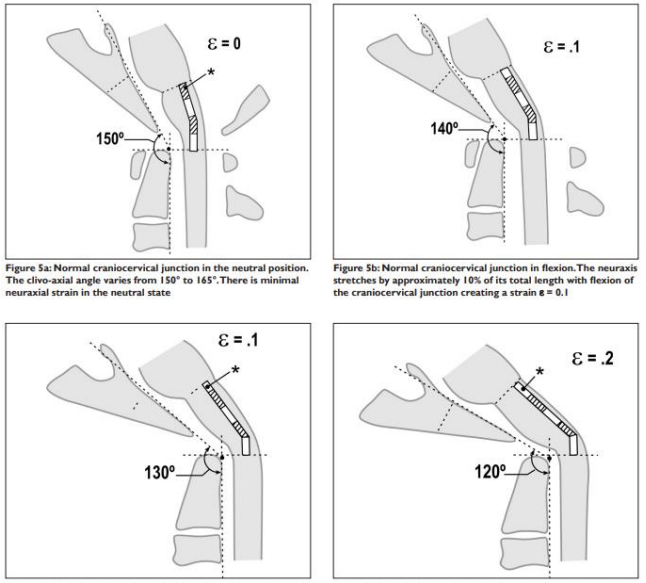
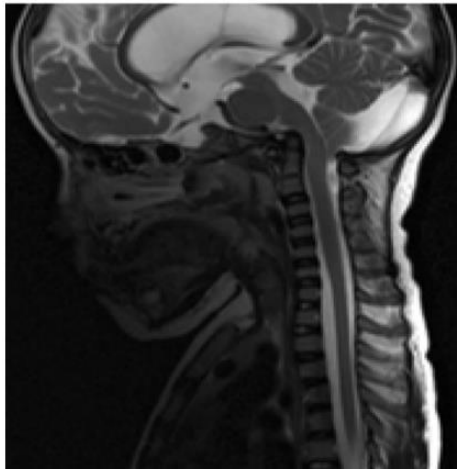


Figure 5a: Normal craniocervical junction in the neutral position. The clivo-axial angle varies from 150° to 165°. There is minimal neuraxial strain in the neutral state

Figure 5b: Normal craniocervical junction in flexion. The neuraxis stretches by approximately 10% of its total length with flexion of the craniocervical junction creating a strain ε = 0.1

direkte Kompression /»stretching» des ZNS

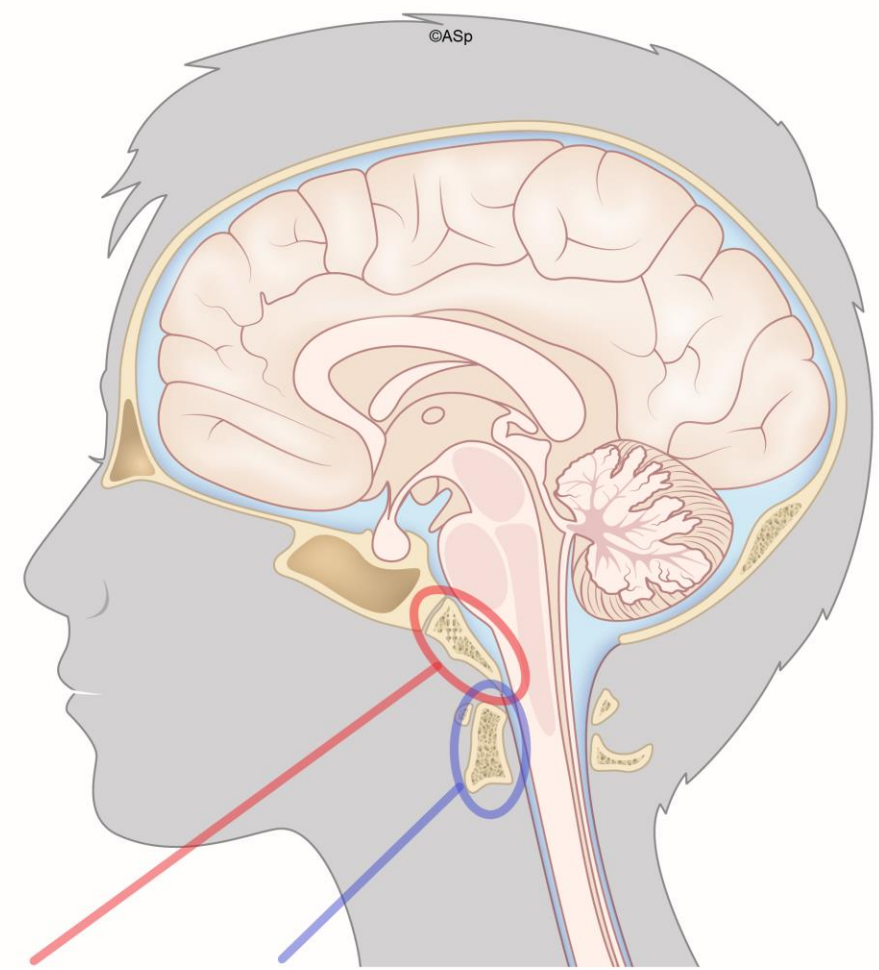


Hydrocephalus (Syrinx)

Henderson FC, Wilson WA, Mott S, et al. Deformative stress associated with an abnormal clivo-axial angle: A finite element analysis. *Surg Neurol Int*. 2010;1:30.
Castelein RM, Hasler C, Helenius I, Ovadia D, Yazici M; EPOS Spine Study Group. Complex spine deformities in young patients with severe osteogenesis imperfecta: current concepts review. J Child Orthop. 2019 Feb 1;13(1):22-32.

Embryologie

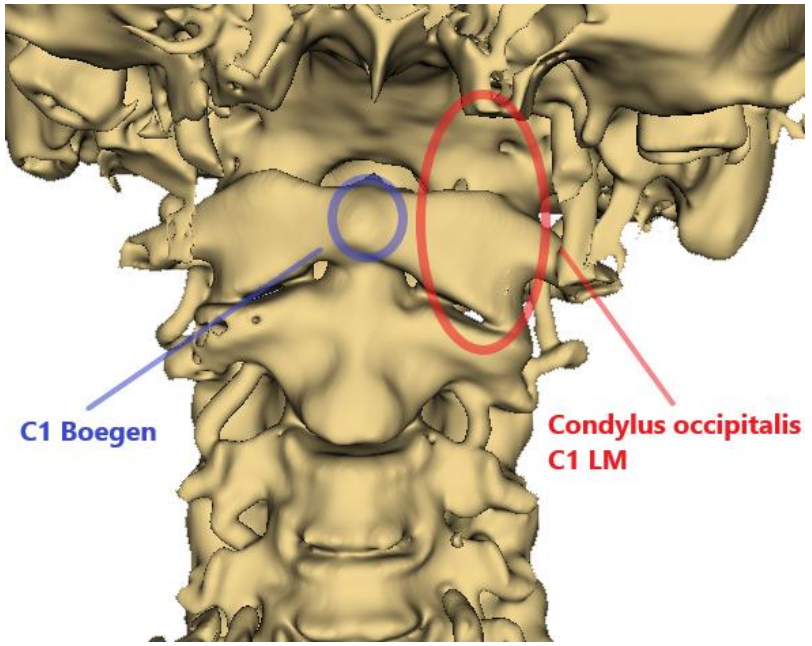
Zentrale Säule



Clivus (Pars basilaris des Os occipitalis)

Odontoid

Laterale Strukturen



Embryologie

Zentrale Säule

- Basilar impression / Invagination
- Bifid Clivus
- Segmentierungsstörung Proatlas Sklerotom
  - Os avis
- Aplasie/Hypoplasie Dens
- Bifid Dens
- Störung der odontoialen Synchondrose
  - Os odontoideum
  - Ossiculum terminale

Laterale Strukturen

- Mobile Clivus (posterior homeotic transformation)
- Akzessorischer Occipital Kondyl
- Occipitocondyäre Hyperplasie
- Pre-Basioccipitaler Bogen
- Atlas Assmiliation (anterior homeotic transformation)
- C1 anterior arch aplasia/hypoplasia
- C1 posterior arch aplasia/hypoplasia
- C1 lateral mass (and anterior arch) aplasia/hypoplasia
- C1 anterior and posterior arch def
- Bifid C1

Diagnose

CT flexion/extension (selten Rotation)
MRI (Tonsillenektopie, Syringomyelie, CSF flow studies, Vaskuläre
Anatomie A. vertebralis/hintere Zirkulation

Basiläre Impression

Def: HWS (Densspitze) innerhalb des Schädels
Radiologische Rkiterien (Chamberlain: >3mm, McGregor >5mm)
2 Typen: Typ 1 primäre Instabilität -> «cranial settling», Typ 2 primäre Schädelbasisanomalie)

Tpye 1

Short clivus causing anterior form of basilar impression

Combined anterior and posterior BI

Platybasia with short clivus BI, TE and syrinx

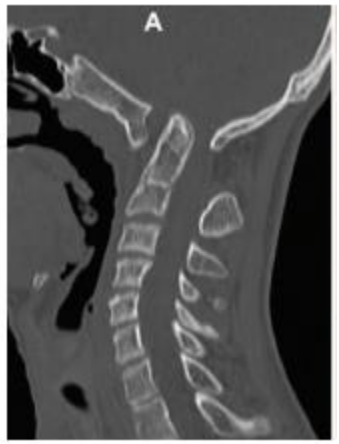
Basiläre Impression

Short clivus causing anterior form of basilar impression

Basilar Impression: Typ I (cranial settling), type II (abnormals skull base anatomy)

Combined anterior and posterior BI

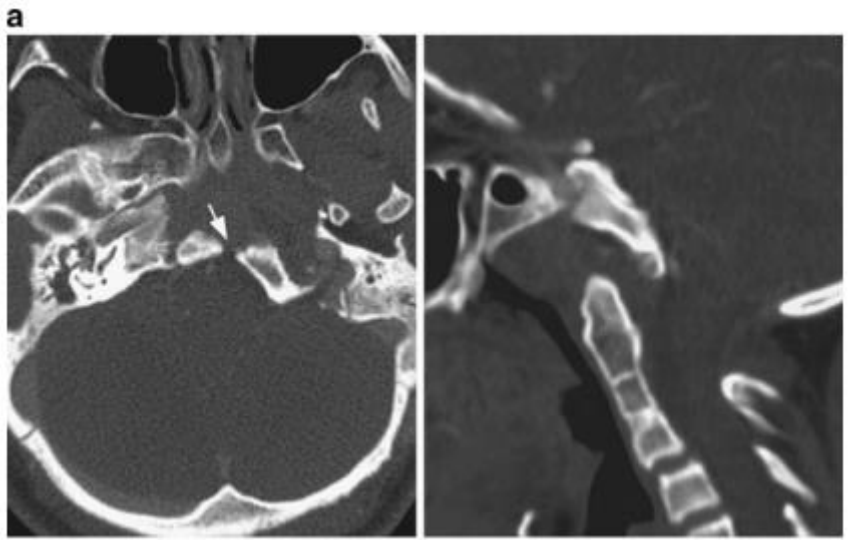
Platybasia with short clivus BI, TE and syrinx



Type I, cranial settling (eg atlas assimilation with C1/2 instabilti)

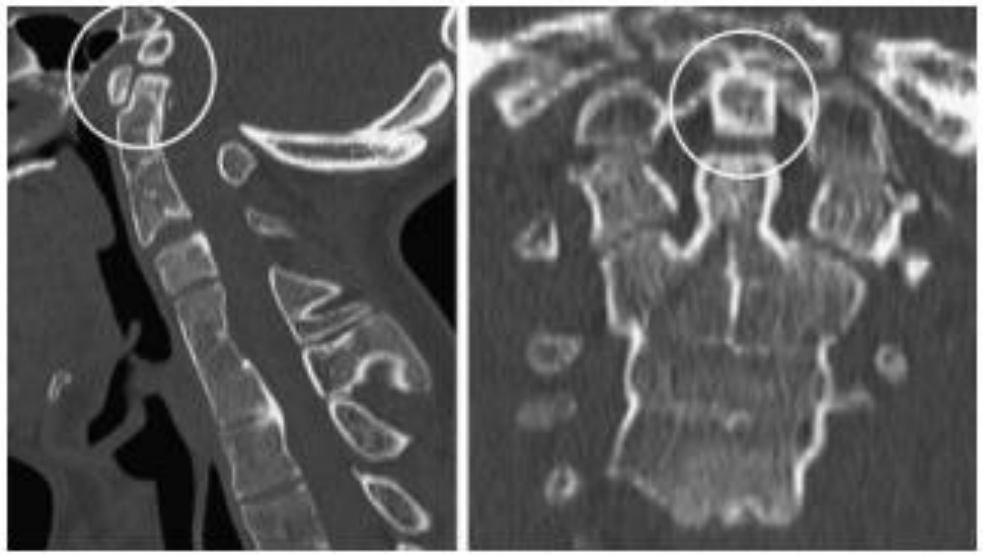
Often outside of syndroms

McGirt MJ, Attenello FJ, Sciubba DM, Gokaslan ZL, Wolinsky JP. Endoscopic transcervical odontoidectomy for pediatric basilar invagination and cranial settling. Report of 4 cases. J Neurosurg Pediatr. 2008 Apr;1(4):337-42. doi: 10.3171/PED/2008/1/4/337. PMID: 18377313.



Bifid clivus

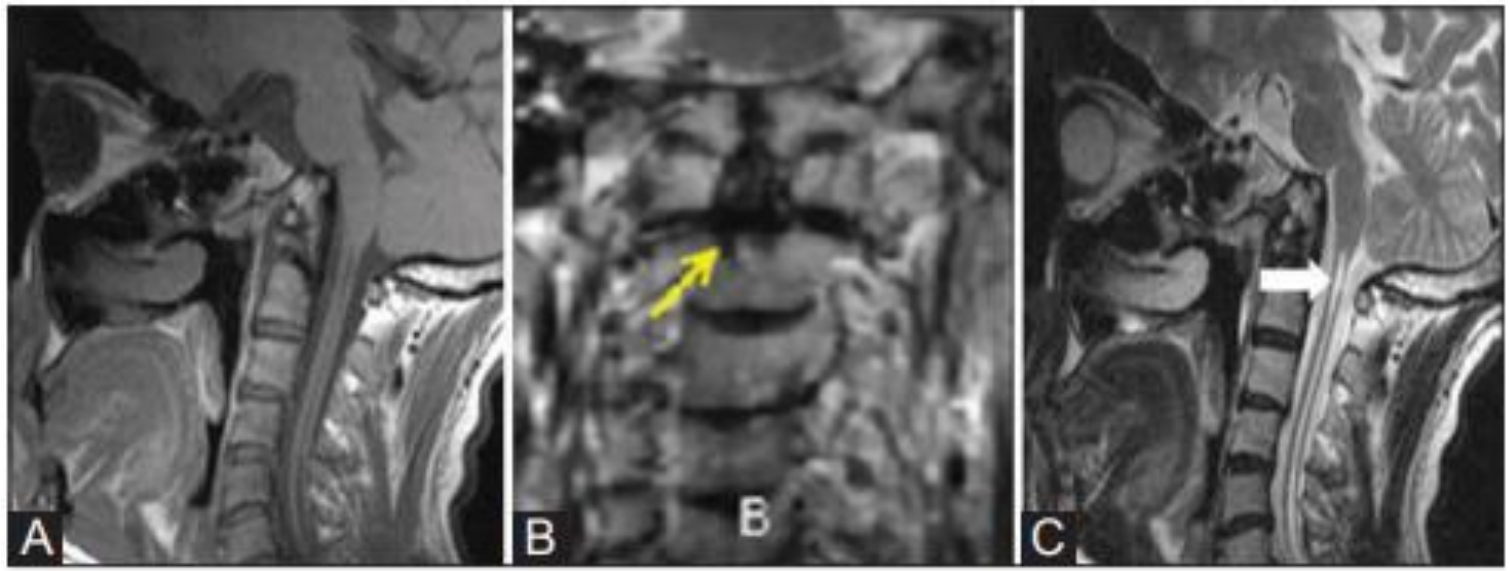
Tectorial membrane anchorage
Might cause C0/C1 instability



Os avis

Might cause C1/C2 instability

Zentrale Säule – Dysgenesie des Odontoid

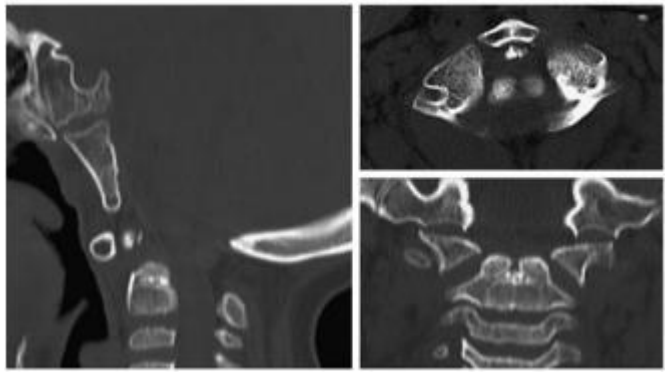


Odontoid aplasia

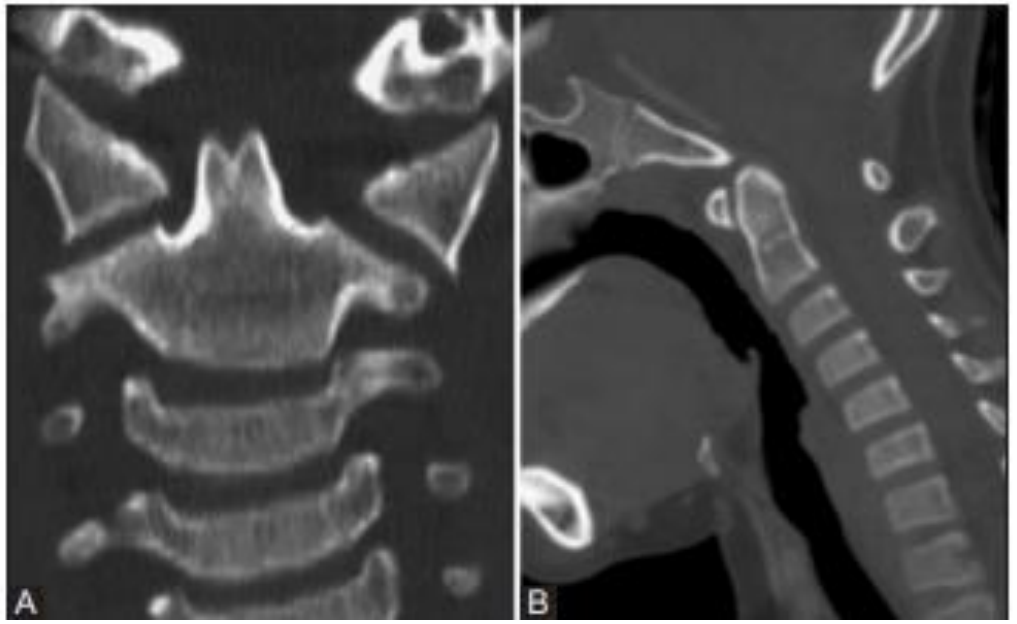
Spondyloepiphyseal, spondylometaphyseal dysplasia
Pure odontoid aplasia, autosomal dominant

Instability C1/C2

Zentrale Säule – Dysgenesie des Odontoid

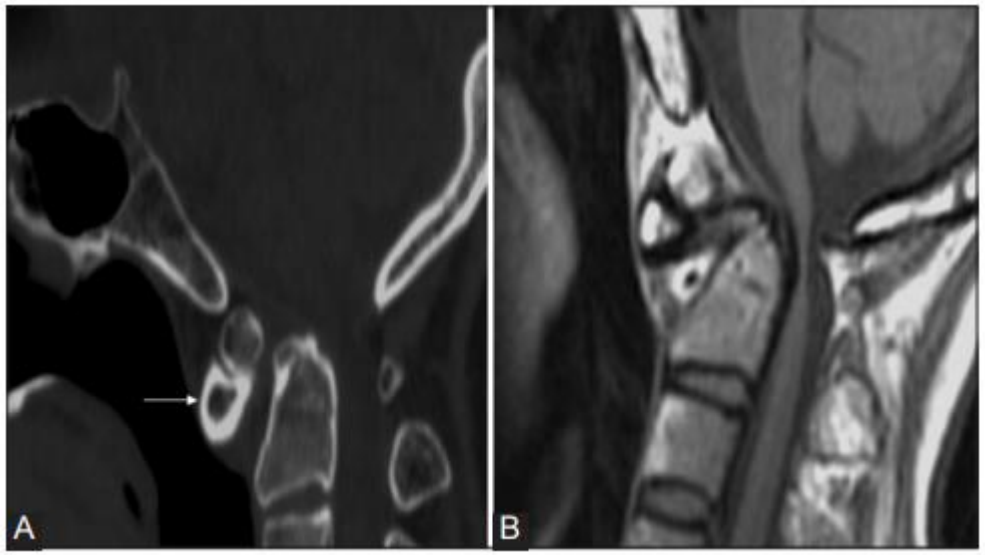


Thompson paper: bifid dens
TAL incompeten
Associated with micrognathia and microstomia



Dens bicornis
Funktionel irrelevatn

Zentrale Säule – Dysgenesie des Odontoid



Os odontoideum
TAL meist nicht intakt
Symptome (transienter Sz und Schwäche bis LCN und rezed. Strokes)

PubmedID 18939918

Mean age at presentation 20.5y (1.5-73y), majority 11-15
Association: 3/78 Down, 1/78 spondyloepiphyseal dysplasia

Other syndromes: Morquio and pseudoachondroplasia

Symptomas: neck pain (64%), neurologic signs 23%

Diagnosis: 56% trauma related

Zentrale Säule – Dysgenesie des Odontoid

Persitent os terminale

TAL meist intakt

Jain N, Verma R, Garga UC, Baruah BP, Jain SK, Bhaskar SN. CT and MR imaging of odontoid abnormalities: A pictorial review. Indian J Radiol Imaging. 2016 Jan-Mar;26(1):108-19.

- Pang D, Thompson DN. Embryology and bony malformations of the craniovertebral junction. *Childs Nerv Syst*. 2011;27(4):523-564. doi:10.1007/s00381-010-1358-9

Platybasia in kids

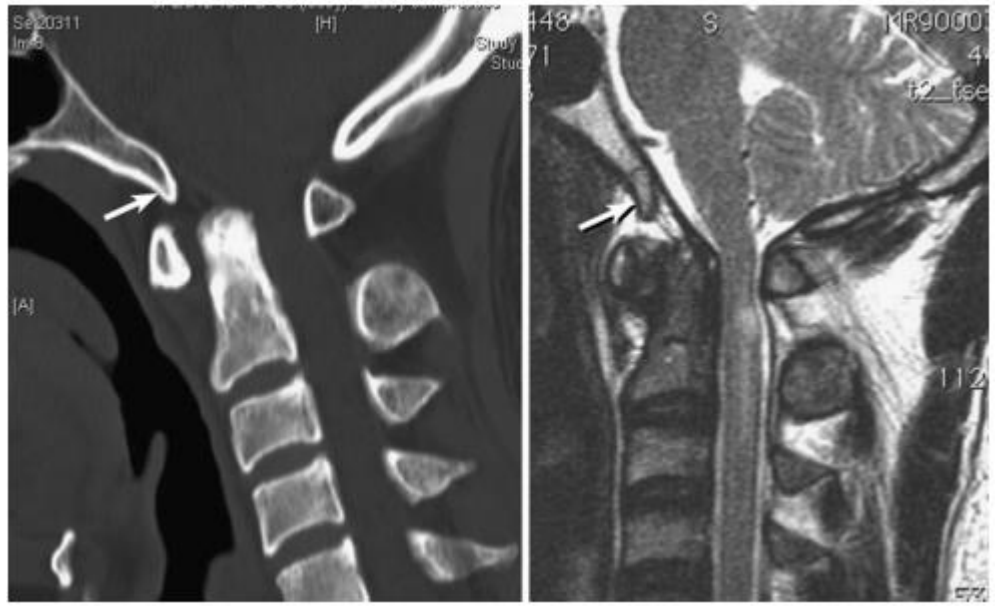
Kann asymptomatisch sein, ohne Assoziation
Osteogenesis imperfecta
Achondroplasia
Down
OI
Morquio

Fibrous dysplasia
Hypopara
osteomalacia

Cranialsettling

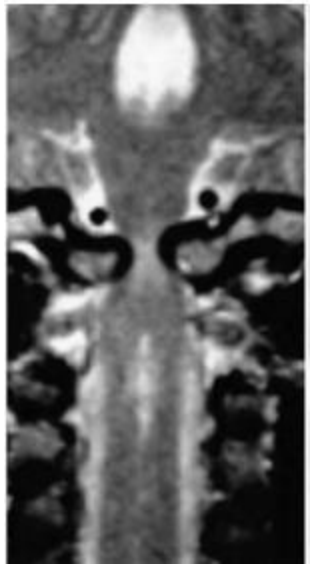
Kann asymptomatisch sein, ohne Assoziation
Osteogenesis imperfecta
Achondroplasia
Down
OI
Morquio

Fibrous dysplasia
Hypopara
osteomalacia

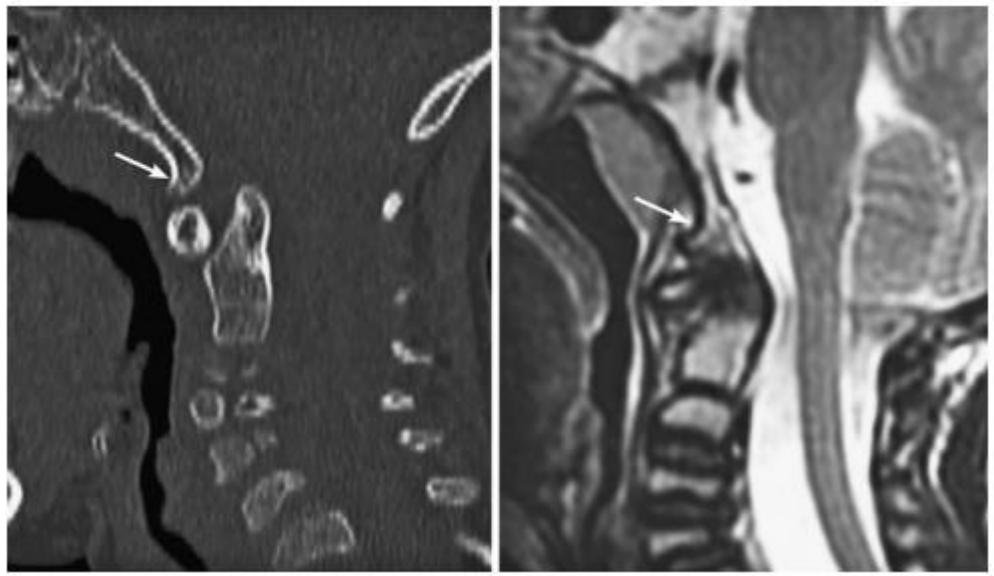


Third occipital condyle

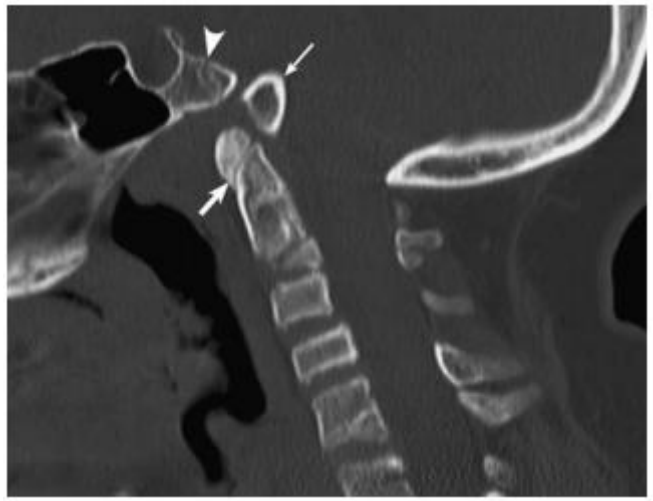
Occipitocondylar hyperplasia



Ohaegbulam C, Woodard EJ, Proctor M. Occipitocondylar hyperplasia: an unusual craniovertebral junction anomaly causing myelopathy. Case report. J Neurosurg. 2005 Oct;103(4 Suppl):379-81. doi: 10.3171/ped.2005.103.4.0379. PMID: 16270692.



Pre-basioccipital arch



Unfused clivus to basiccipital

- Henderson, F.C., Henderson, F.C., Wilson, W.A. *et al.* Utility of the clivo-axial angle in assessing brainstem deformity: pilot study and literature review. *Neurosurg Rev* **41,** 149–163 (2018). https://doi.org/10.1007/s10143-017-0830-3

EDS

- Mourad F, Giovannico G, Maselli F, Bonetti F, Fernández de las Peñas C, Dunning J. Basilar impression presenting as intermittent mechanical neck pain: a rare case report. BMC Musculoskelet Disord. 2016 Jan 11;17:7. doi: 10.1186/s12891-015-0847-0. PMID: 26754441; PMCID: PMC4707768.


SC SAG T2 FRFSE
H
A
W1317 / C658
TE: 104.244003296898
TR: 1960
Slice pos: -9.5 ST: 4.0mm
Position: HFS
102 IMA 5

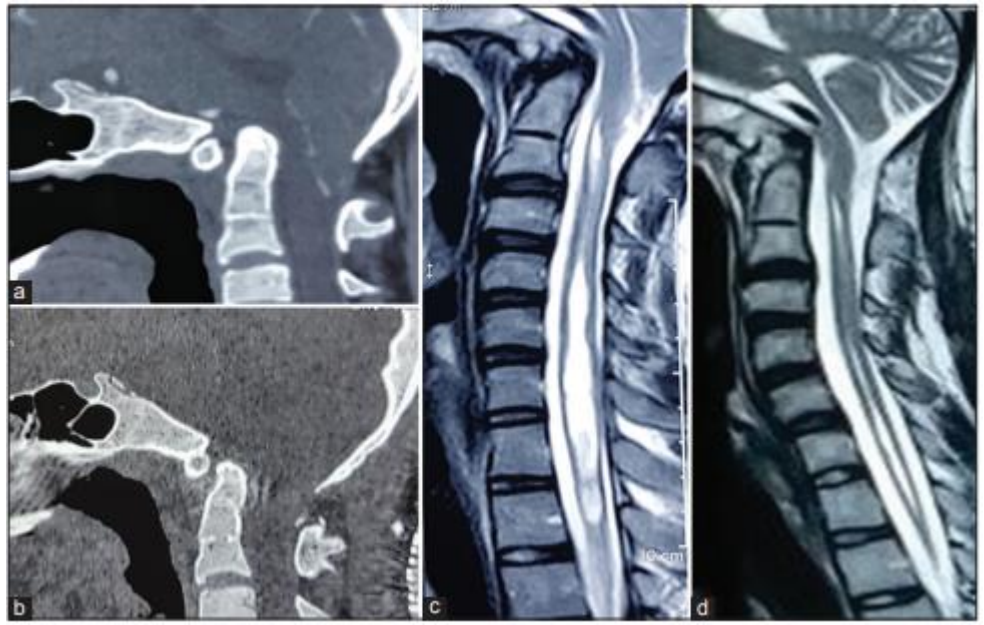

SC CORT2 FRFSE
HP
R
W883 / C441
TE: 104.1760025032441
TR: 2260
Slice pos: 19.8 ST: 4.0mm
Position: HFS
105 IMA 2
Zoom factor: x0.97

- Cobanoglu M, Bauer JM, Campbell JW, Shah SA. Basilar impression in osteogenesis imperfecta treated with staged halo traction and posterior decompression with short-segment fusion. *J Craniovertebr Junction Spine*. 2018;9(3):212-215. doi:10.4103/jcvjs.JCVJS_63_18

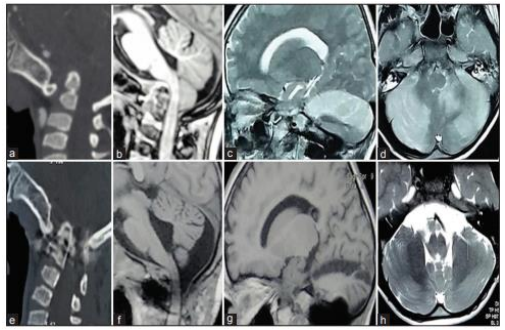

g


b

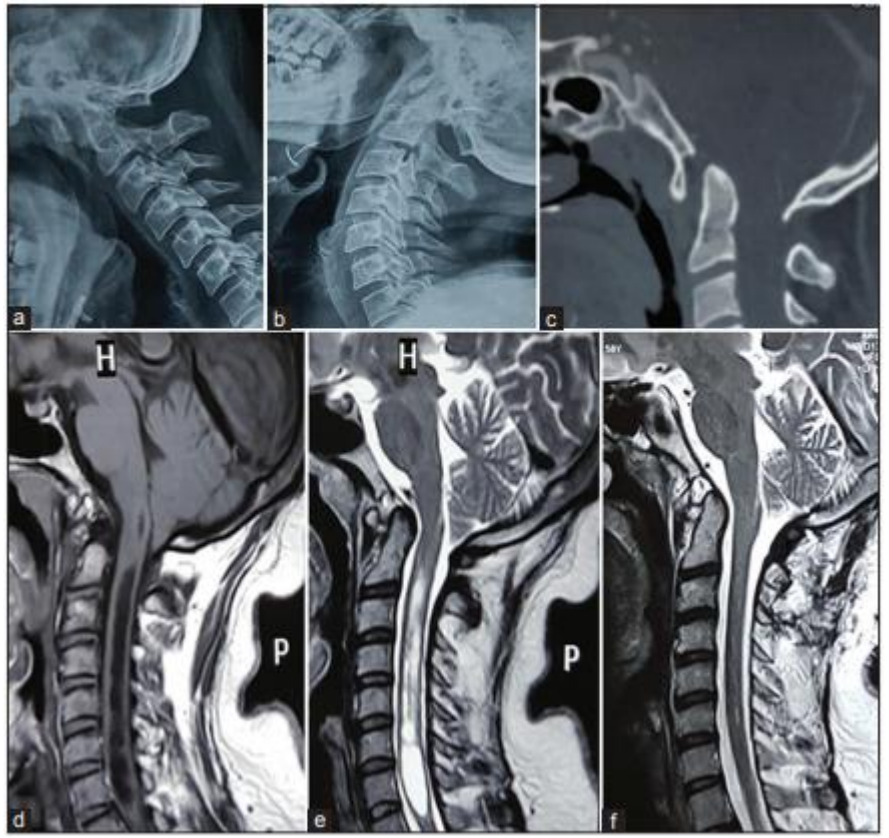
- Salunke P, Karthigeyan M, Malik P. Foramen magnum decompression without bone removal: C1-C2 posterior fixation for Chiari with congenital atlantoaxial dislocation/basilar invagination. Surg Neurol Int. 2019 Mar 26;10:38. doi: 10.25259/SNI-38-2019. PMID: 31528376; PMCID: PMC6743679.



C1 assimilation, C1/C2 instability, less platybasia, CXA

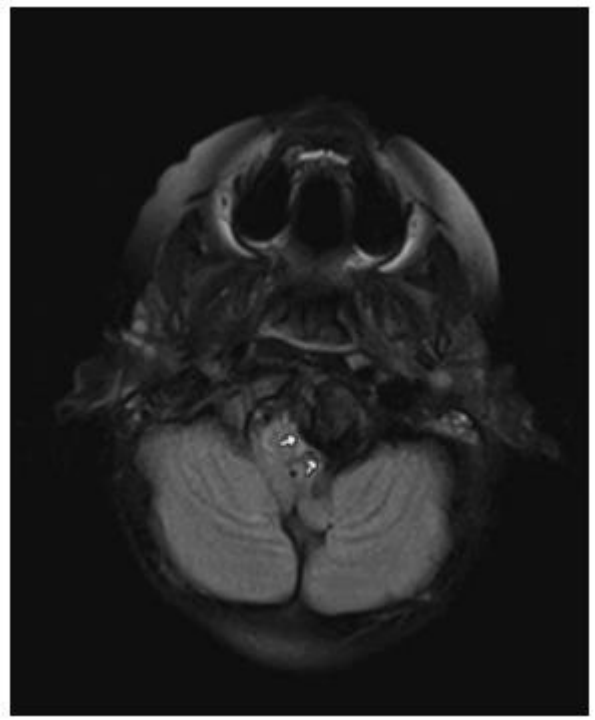


Atlas assimilation and C1/C2 instab



Atlas assimilation and C1/C2 instab

- Nagy L, Ray C. Spontaneously improving occipitocondylar hyperplasia: a case report. J Neurol Surg Rep. 2014 Aug;75(1):e141-3. doi: 10.1055/s-0034-1376426. Epub 2014 May 28. PMID: 25083374; PMCID: PMC4110150.



Hyperplatsic occipital condyle

- Ji W, Liu X, Huang W, Huang Z, Chen J, Zhu Q, Wu Z. Clival Screw Placement in Patient with atlas assimilation: A CT-based feasibility study. Sci Rep. 2016 Aug 19;6:31648. doi: 10.1038/srep31648. PMID: 27539005; PMCID: PMC4990935.

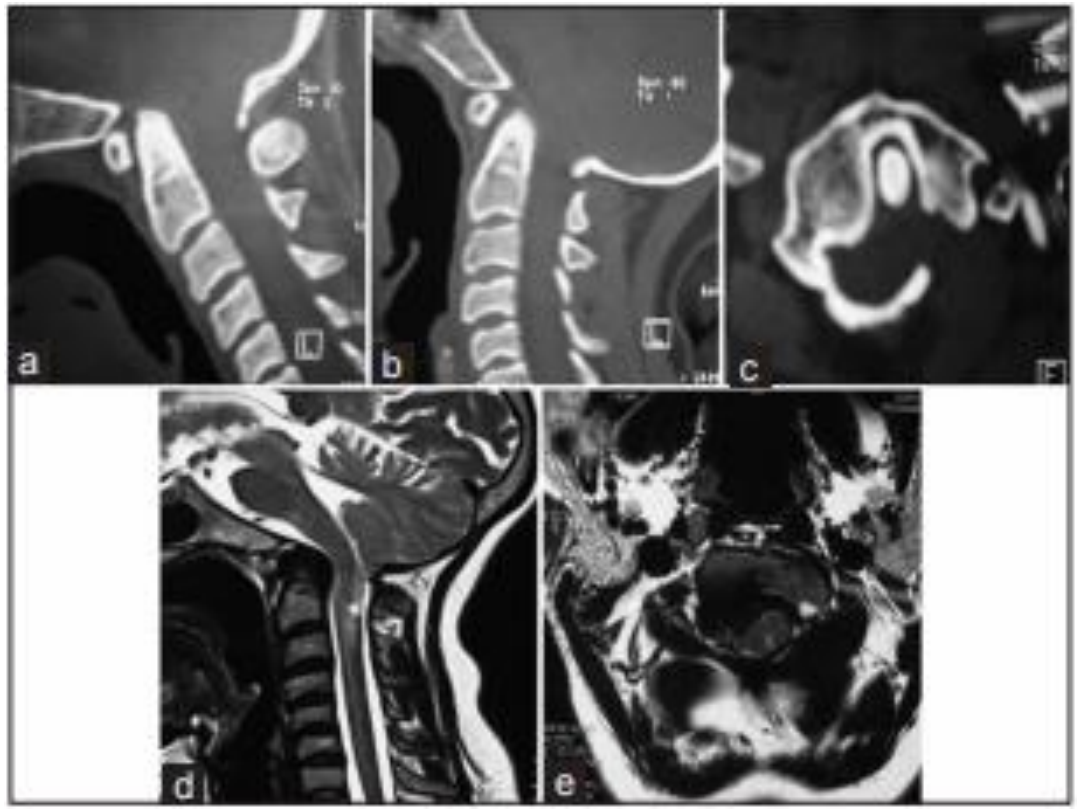
Menezes AH, Dlouhy BJ. Atlas assimilation: spectrum of associated radiographic abnormalities, clinical presentation, and management in children below 10 years. Childs Nerv Syst. 2020 May;36(5):975-985. doi: 10.1007/s00381-019-04488-3. Epub 2020 Jan 4. PMID: 31901967.

Atlas assimilation

In the pediatric population: 12% C1/C2 dislocation, 42% tonsillar ectopia

Associated findings: C2/C3 fusion 74%, CMI (42%), condylar hypolasp (31%),

- Das KK, Mehrotra A, Sahu RN, Srivastava AK, Jaiswal AK, Behari S. Unilateral lateral mass hypertrophy: An extremely rare congenital anomaly of atlas. *J Craniovertebr Junction Spine*. 2013;4(2):73-75. doi:10.4103/0974-8237.128534



C1 lateral mass hyperplasia

- N V A, Avinash M, K S S, Shetty AP, Kanna RM, Rajasekaran S. Congenital Osseous Anomalies of the Cervical Spine: Occurrence, Morphological Characteristics, Embryological Basis and Clinical Significance: A Computed Tomography Based Study. *Asian Spine J*. 2019;13(4):535-543. Published 2019 Mar 14. doi:10.31616/asj.2018.0260

Atlas assimilation

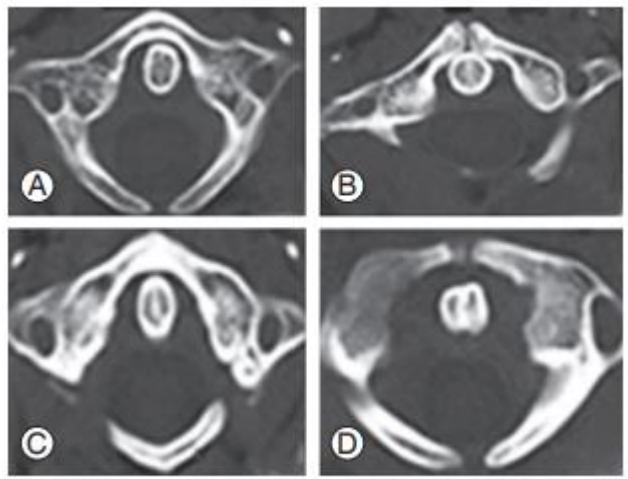


Fig. 5. Various patterns of arch defects of the atlas. (A) Midline; (B) unilateral; (C) bilateral; (D) anterior and posterior (spondyloschisis).

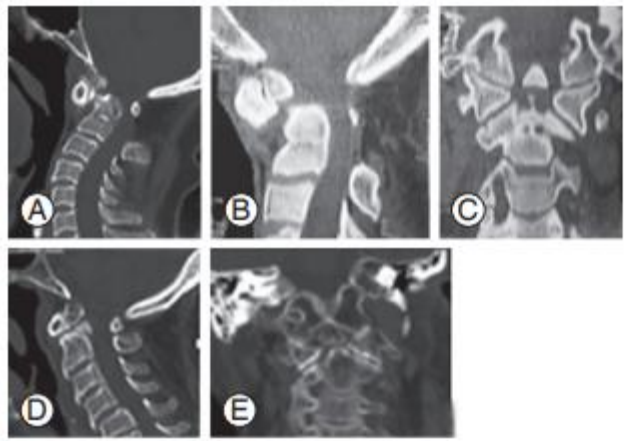


Fig. 6. (A) Dystopic os odontoideum with the odontoid fragment fused to the basion. (B, C) Sagittal and coronal images show orthotopic os odontoideum with round margins and wider clear space between the ossicle and axis to be differentiated from nonunion odontoid (D, E).

OP Technik – posterior reduction

Fixiertes cranial semi-rigide;

Posteriore instrumentierte Fusion; Stäbe fixiert an HWS, frei Occiput -> Distraktion in sagittaler Ebene und Kompression axialer Ebene

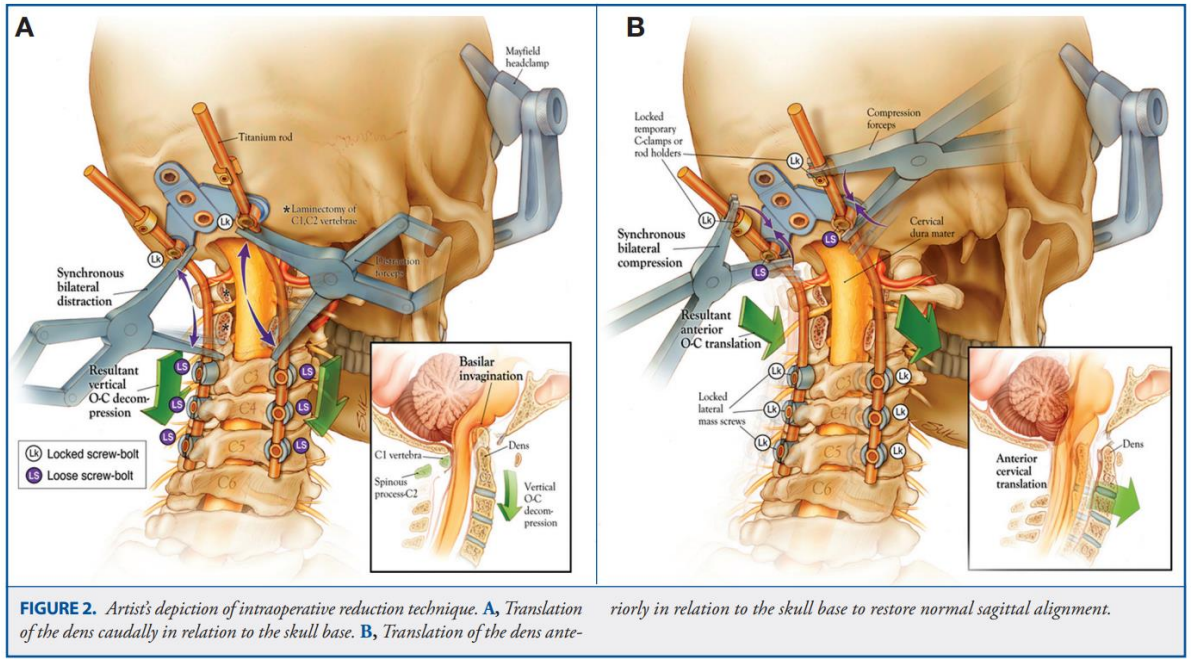


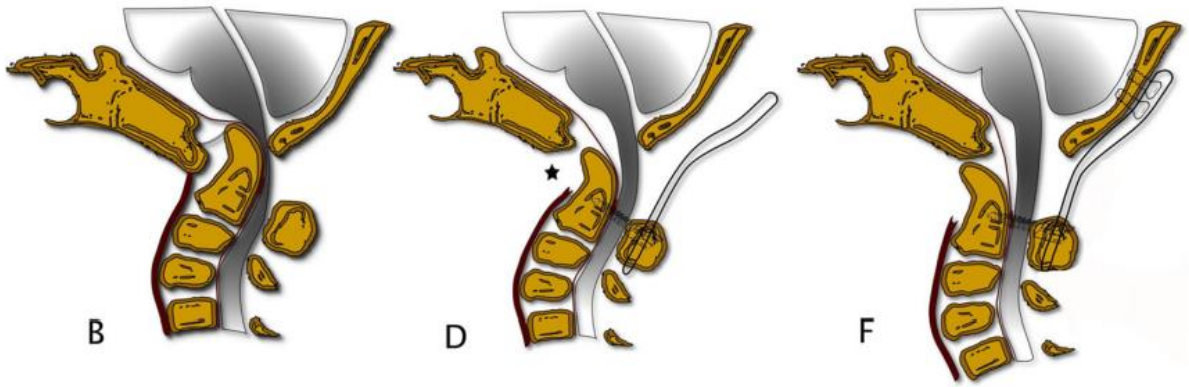
FIGURE 2. *Artist's depiction of intraoperative reduction technique.* **A,** *Translation of the dens caudally in relation to the skull base.* **B,** *Translation of the dens anteriorly in relation to the skull base to restore normal sagittal alignment.*

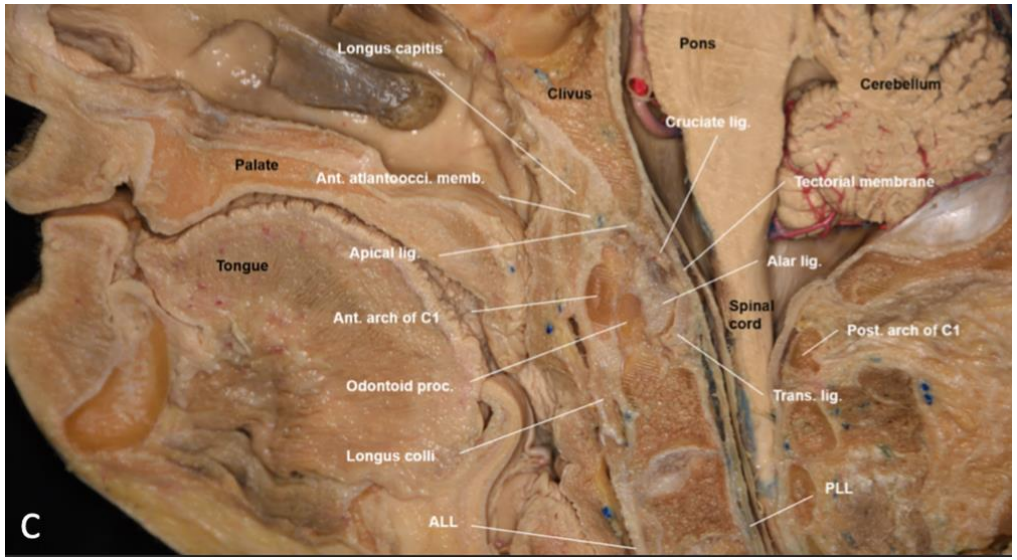
Hsu Wesley, MD, Hasan A. Zaidi, MD, Ian Suk, BSc, BMC, Ziya L. Gokaslan, MD, Jean-Paul Wolinsky, MD, A New Technique for Intraoperative Reduction of Occipitocervical Instability, *Operative Neurosurgery*, Volume 66, Issue suppl_2, June 2010, Pages ons319–ons324,

OP Technik – anterior release

Fixiertes cranial settling

Transsektion von M. longus colli und vorderes Längsband -> Mobilisation Kopf von der Wirbelsäule –> Fusion





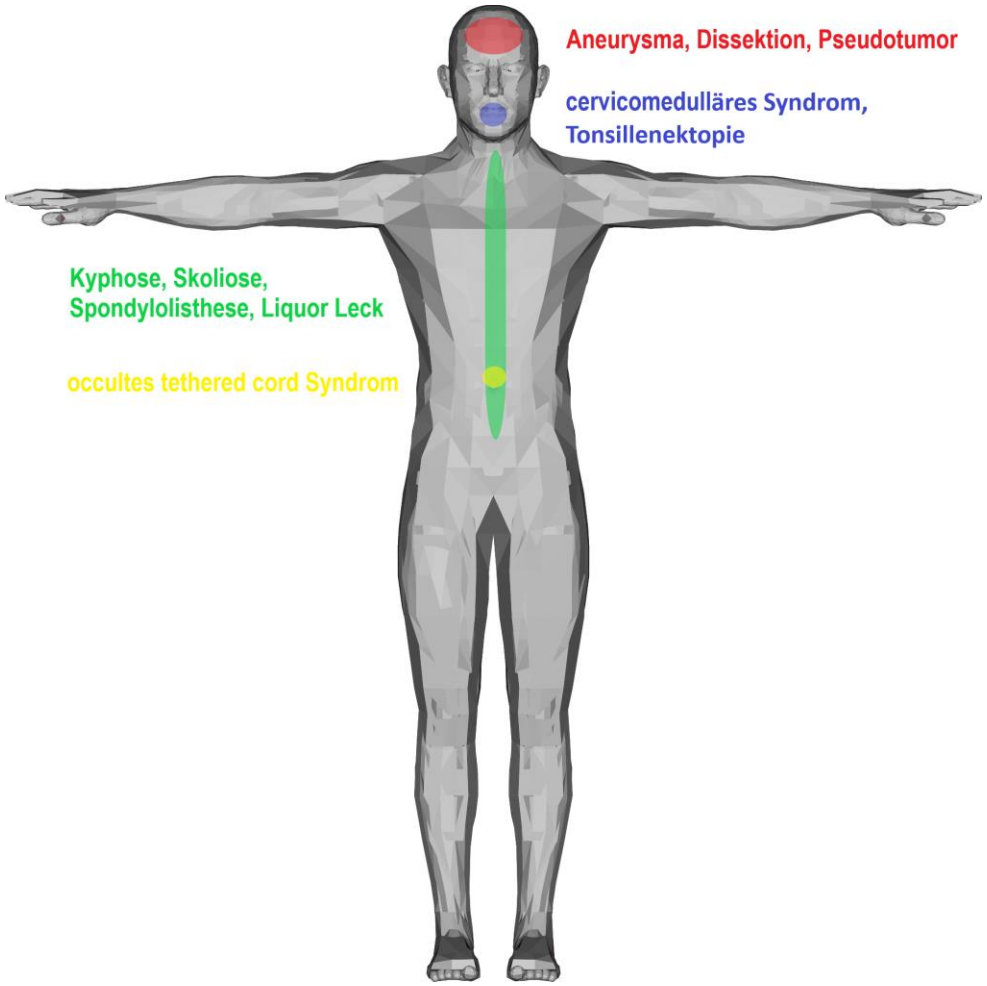
Laheri, V., Chaudhary, K., Rathod, A. *et al.* Anterior transoral atlantoaxial release and posterior instrumented fusion for irreducible congenital basilar invagination. *Eur Spine J* **24,** 2977–2985 (2015). https://doi.org/10.1007/s00586-015-3836-y

Stabilisierung Occiput – C2

Dekompression
-Posterior fossa decompression
-

Geschlossene oder offene Reposition
- präop. Axiale Traktion mit Crown-Halo
- intraop. «Kopfreposition»
- intraop. Reduktionsmaneuver

Neurochirurgische Manifestationen von EDS



Pathologie	EDS Subtyp
Aneurysma, Dissektion	klassisch, hypermobil, vaskulär
Pseudotumor cerebri	hypermobil
Cervicomedulläres Syndrom / Tonsillenektopie	klassisch, hypermobil
Skoliose / Kyphoskoliose	klassisch, hypermobil, vaskulär, kyphoskoliotisch
Spontanes Liquor Leck	hypermobil
Occultes Tethered Cord Syndrom	hypermobil

Neurochirurgische Manifestationen von EDS

Neurochirurgische Entitäten sind

- NICHT pathognomonisch in Bezug auf EDS
- Wissen unter Kliniker oft wenig ausgeprägt
- Diagnose oft fordernd da: «Routine» Bildgebung oft nicht adäquat
- Vieles erst teilweise «erforscht», insbesondere Zusammenhänge oft noch nicht klar

Ziel des Vortrages

- Wissen/»Awareness» unter Ärzten, welche EDS Patienten behandeln zu optimieren
- evtl. kollaborativ neues Wissen zu schaffen

Kranielles Aneurysma, Dissektion, AV Fistel

Aneurysma

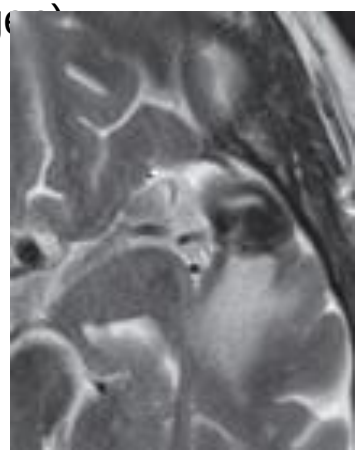
Inzidenz: 12% aller EDS Patienten (klassisch, hypermobil, vaskulär)

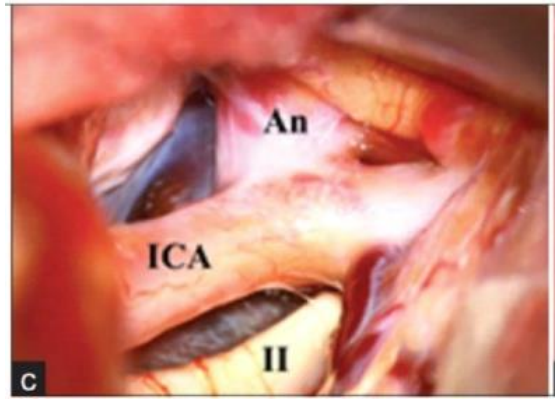
Klinische Korrelate:

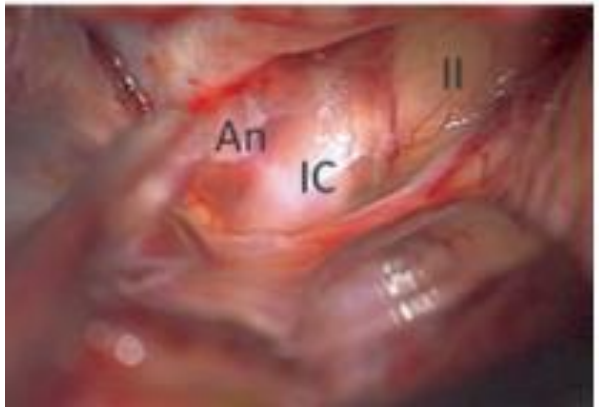
nicht rupturiert und asymptomatisch

nicht rupturiert und symptomatisch (TIA, Schlaganfall, Kopfschmerz, Epi Anfall, Hirnnervenlähmung)

rupturiert







Onofrj V, Tampieri D, Cianfoni A, Ventura E. Peri-Aneurysmal Brain Edema in Native and Treated Aneurysms: The Role of Thrombosis [published correction appears in Neurointervention. 2021 Apr 05;:]. *Neurointervention*. 2021;16(1):70-77.

Kranielles Aneurysma, Dissektion, AV Fistel

Aneurysma

Aneurysma Ruptur ist nachwievor ein «life-changing» Event

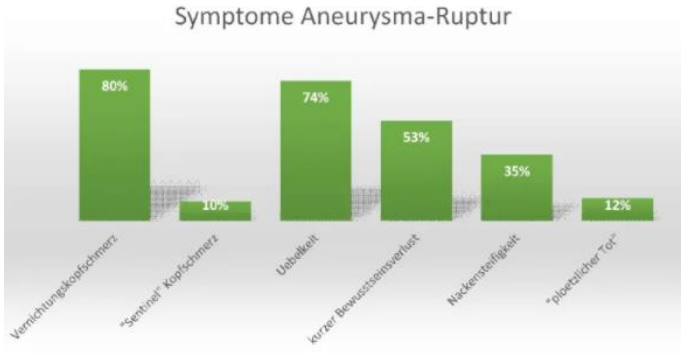
Screening von gefährdeten EDS mittels MR-A; Aneurysmen bilden sich NEU zeitlebens

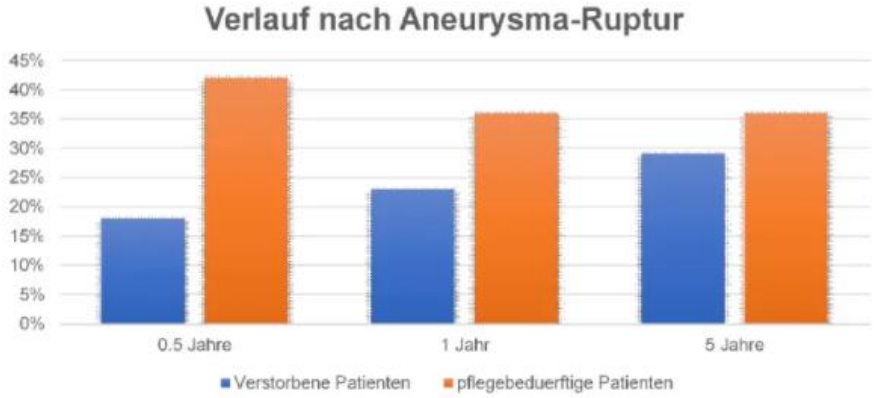
Anbindung an Neurovaskuläres Zentrum

Früherkennung von undiagnostizierten oder «aktiven» Aneurysmen (Epi Anfall, CVI, CN Ausfälle)

Früherkennung “Sentinel” Blutung

V.a. Ruptur: CT vs LP vs MRI





Connolly ES Jr, Rabinstein AA, Carhuapoma JR, Derdeyn CP, Dion J, Higashida RT, Hoh BL, Kirkness CJ, Naidech AM, Ogilvy CS, Patel AB, Thompson BG, Vespa P; American Heart Association Stroke Council; Council on Cardiovascular Radiology and Intervention; Council on Cardiovascular Nursing; Council on Cardiovascular Surgery and Anesthesia; Council on Clinical Cardiology. Guidelines for the management of aneurysmal subarachnoid hemorrhage: a guideline for healthcare professionals from the American Heart Association/american Stroke Association. Stroke. 2012 Jun;43(6):1711-37.

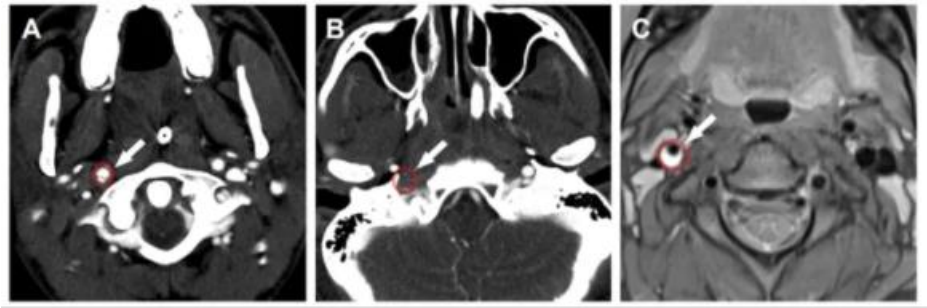
Kranielles Aneurysma, Dissektion, AV Fistel

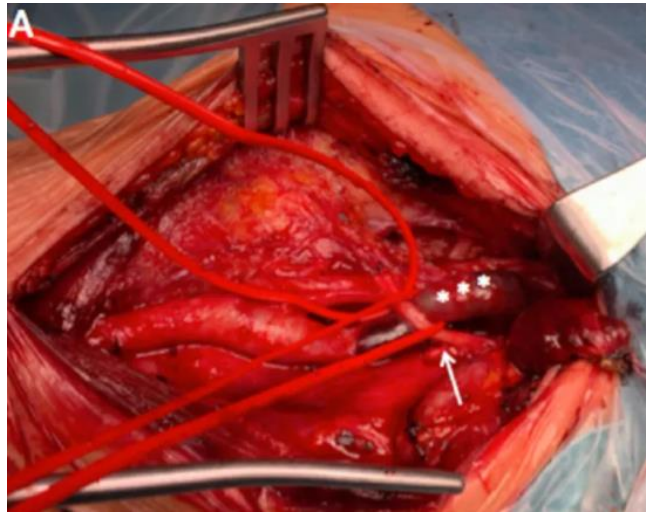
Dissektion (Karotis, Vertebralis)

klassisch, hypermobil, vaskulärer Subtype

Führt oft/meist zu Infarkt

Behandlung: Behandlung stroke, Plättchenhemmung zur stroke Prävention, endovascular oder selten chirurgischer bypass

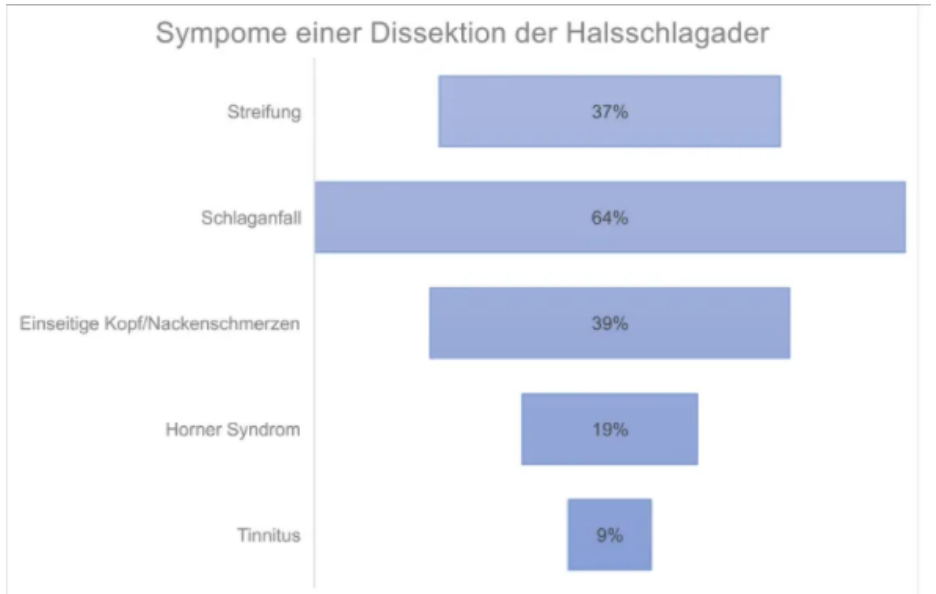




Wu, Yongjun, et al. "Predisposing factors and radiological features in patients with internal carotid artery dissection or vertebral artery dissection." *BMC neurology* 20.1 (2020): 1-8.
Schwartz, Neil E., et al. "Clinical and radiographic natural history of cervical artery dissections." Journal of stroke and cerebrovascular diseases 18.6 (2009): 416-423.

Kranielles Aneurysma, Dissektion, AV Fistel

Dissektion (Karotis, Vertebralis)



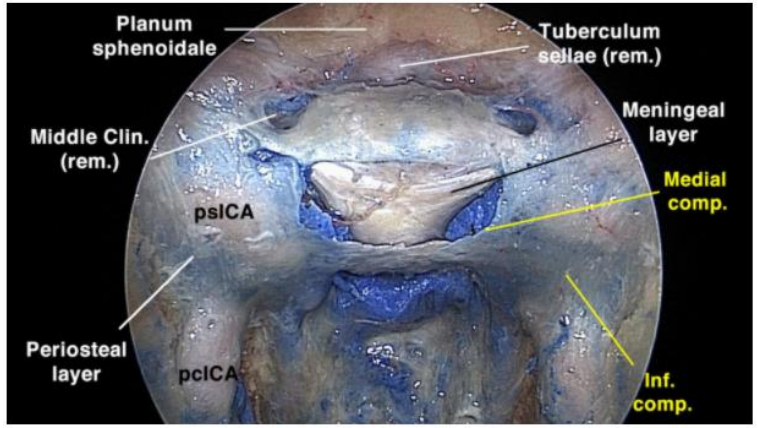
Biousse, Valérie, et al. "Time course of symptoms in extracranial carotid artery dissections: a series of 80 patients." Stroke 26.2 (1995): 235-239.

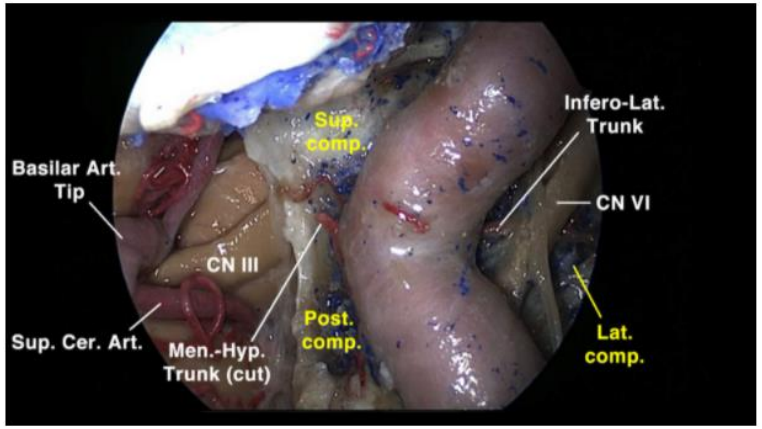
Kranielles Aneurysma, Dissektion, AV Fistel

AV Fisteln

vaskulärer Subtype

In fast allen Fällen Karotis-Sinus cavernosus Fistel





Adham S, Trystram D, Albuisson J, et al. Pathophysiology of carotid-cavernous fistulas in vascular Ehlers-Danlos syndrome: a retrospective cohort and comprehensive review. *Orphanet J Rare Dis*. 2018;13(1):100. Published 2018 Jun 25. doi:10.1186/s13023-018-0842-2

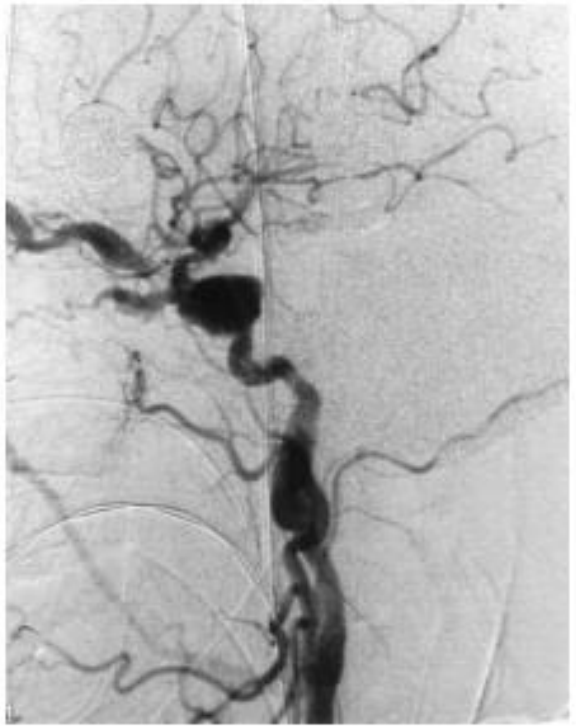
Kranielles Aneurysma, Dissektion, AV Fistel

AV Fisteln

Unterschiedliche Schweregrade, je nach Shuntvolumen

Bei EDS meist geringes Volumen

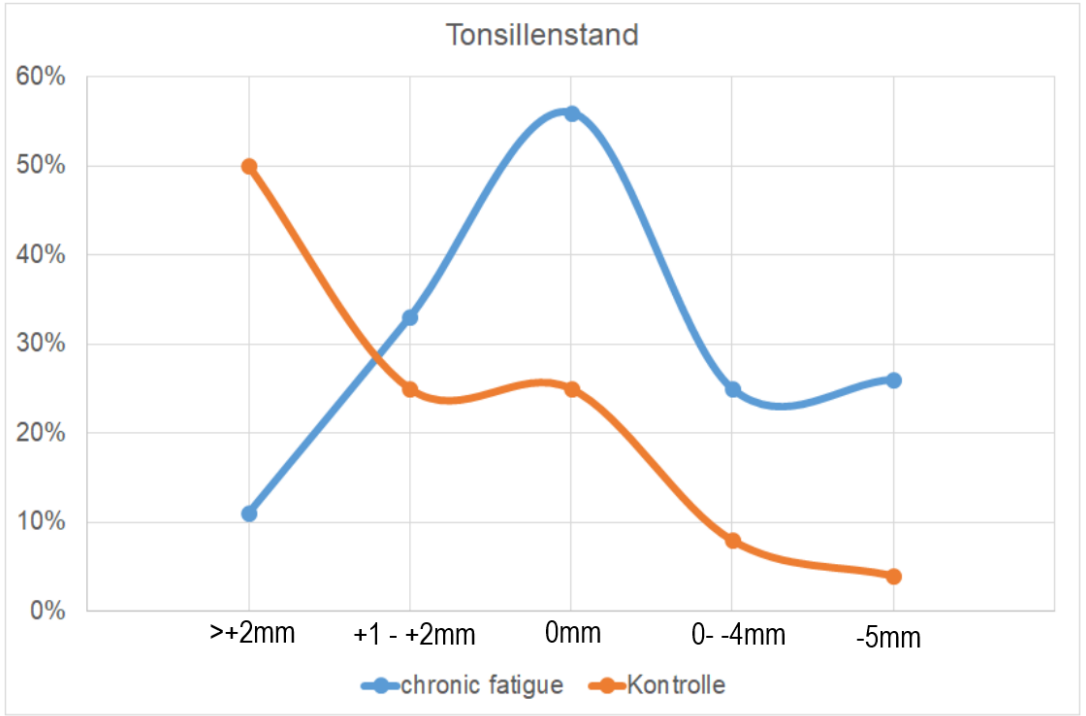
Beschwerden der Augen	*neurologische Beschwerden*	*andere Beschwerden*
Schmerzen Schwellung Rötung Doppelbilder	Sprachstörungen Verwirrtheit	Kopfschmerzen Gesichtsschmerzen

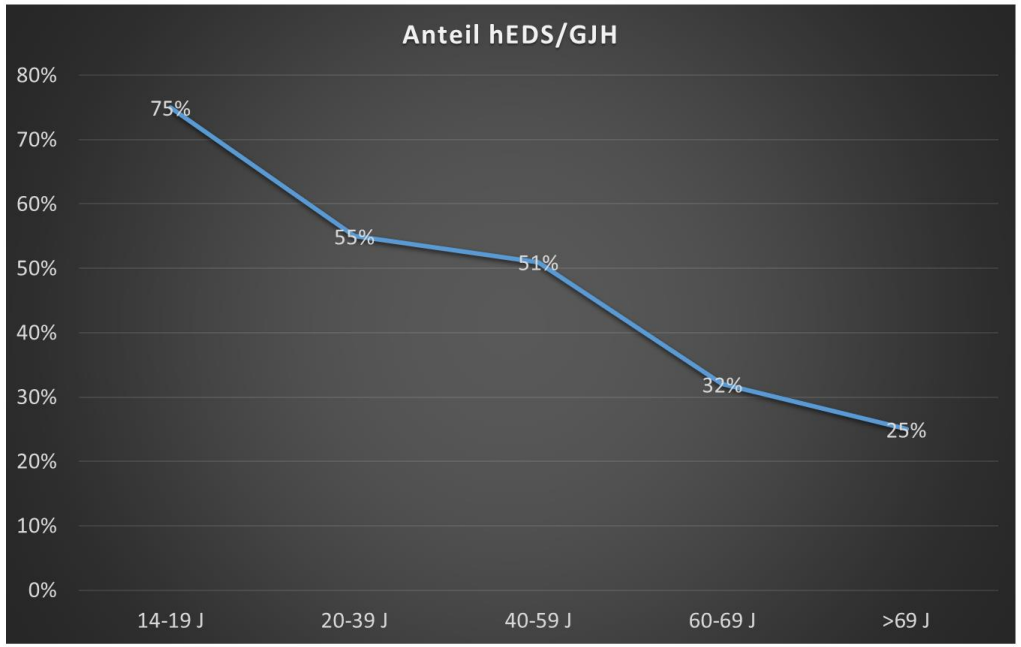


Henderson AD, Miller NR. Carotid-cavernous fistula: current concepts in aetiology, investigation, and management. Eye (Lond). 2018 Feb;32(2):164-172. doi:

Pseudotumor cerebri

hEDS
Kognitive Störungen («brain fog»), ggf.
Sehstörungen
Kann sekundäre Tonsillenektopie bewirken



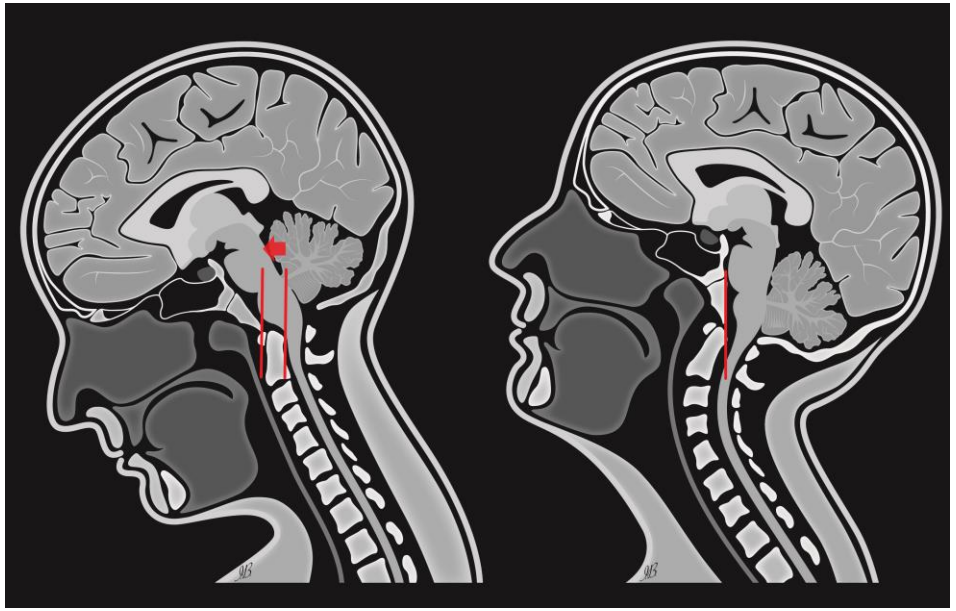


Bragée B, Michos A, Drum B, Fahlgren M, Szulkin R, Bertilson BC. Signs of Intracranial Hypertension, Hypermobility, and Craniocervical Obstructions in Patients With Myalgic Encephalomyelitis/Chronic Fatigue Syndrome. *Front Neurol*. 2020;11:828.

CCI – Cervicomedulläres Syndrom

Pathophysiologie

- ligamentous laxity → subluxation C0/C1 (“odontoid pivoting”)



etiology

- hypermobile EDS and classical EDS
- Marfan syndrome

Background – cervicomedullary syndrome

symptoms

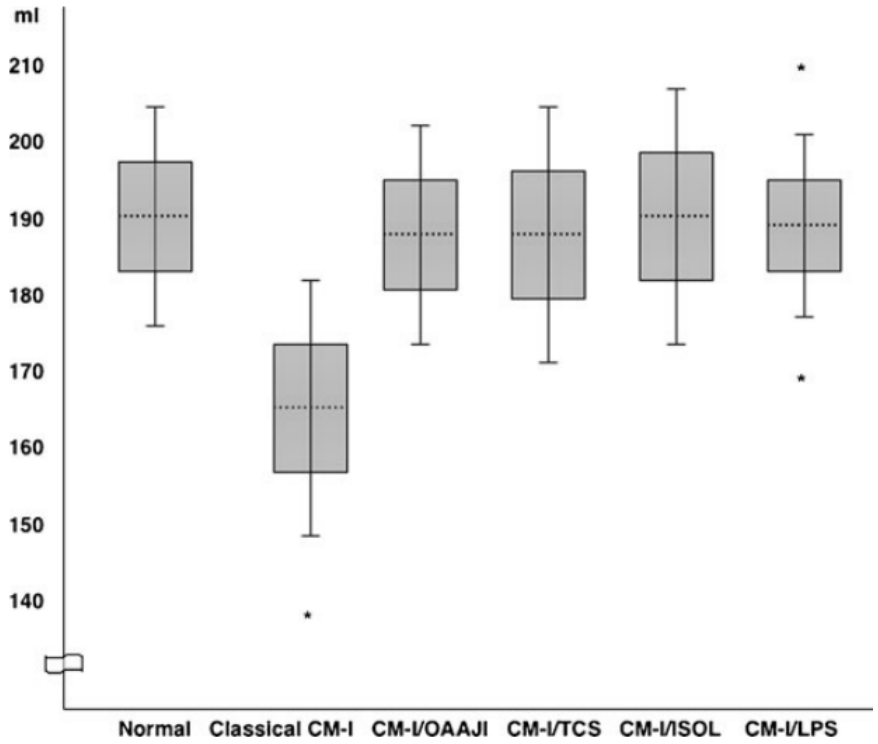
- neck / head pain, nausea, vertigo
- lower cranial nerve dysfunction: dysphagia, throat pain, double vision, nystagmus, tinnitus
- vegetative symptoms (POTS, Raynaud syndrome)

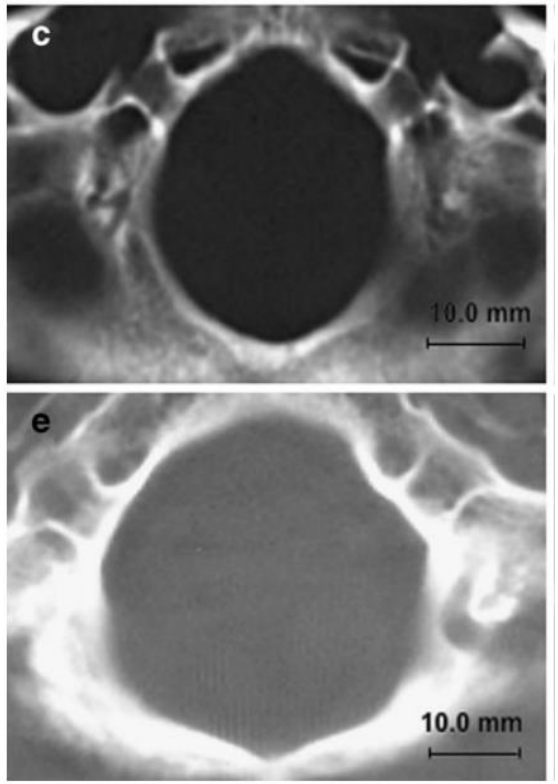
Milhorat TH, Bolognese PA, Nishikawa M, McDonnell NB, Francomano CA. Syndrome of occipitoatlantoaxial hypermobility, cranial settling, and chiari malformation type I in patients with hereditary disorders of connective tissue. J Neurosurg Spine. 2007 Dec;7(6):601-9. doi: 10.3171/SPI-07/12/601. PMID: 18074684.

Diagnostic distinctions of CM-I in patients with and without HDCT

Variable	Patient Group (%)		
	HDCT/CM-I	CM-I	p Value
total no. of cases	357	2456	
symptoms & signs			
nausea	239 (67)	343 (14)	<0.001
dysphagia	246 (69)	639 (26)	<0.001
throat tightness	171 (48)	221 (9)	<0.001
shortness of breath	96 (27)	98 (4)	<0.001
sleep apnea	110 (31)	270 (11)	<0.001
palpitations	218 (61)	514 (21)	<0.001
facial pain	89 (25)	172 (7)	<0.001
double vision	93 (26)	123 (5)	<0.001
syncope	50 (14)	25 (1)	<0.001
diagnostic findings			
Lhermitte sign	246 (69)	74 (3)	<0.001
Raynaud phenomenon	136 (38)	170 (7)	<0.001
downward nystagmus	54 (15)	73 (3)	<0.001
dizziness w/ head turning	133 (37)	196 (8)	<0.001
orthostatic hypotension*	74 (21)	49 (2)	<0.001
postural orthostatic tachycardia syndrome*	92 (26)	123 (5)	<0.001
supraventricular tachycardia†	114 (32)	269 (11)	<0.001
mitral valve prolapse‡	174 (49)	74 (3)	<0.001
mitral valve regurgitation‡	35 (10)	49 (2)	<0.001
radiographic findings			
retroodontoid pannus w/ basilar impression	253 (71)	270 (11)	<0.001
cervical disc disease	207 (58)	295 (12)	<0.001
cervical spine subluxation	67 (19)	48 (2)	<0.001
scoliosis	175 (49)	539 (22)	<0.001
temporomandibular joint disease	231 (65)	419 (17)	<0.001
oropharynx hypoplasia	158 (44)	50 (2)	<0.001
hypertrophy of styloid process	114 (32)	244 (10)	<0.001
mean KPS score§	67.5 ± 3.9	76.8 ± 3.4	<0.001

Milhorat, T.H., Nishikawa, M., Kula, R.W. *et al.* Mechanisms of cerebellar tonsil herniation in patients with Chiari malformations as guide to clinical management. *Acta Neurochir* **152,** 1117–1127 (2010). https://doi.org/10.1007/s00701-010-0636-3

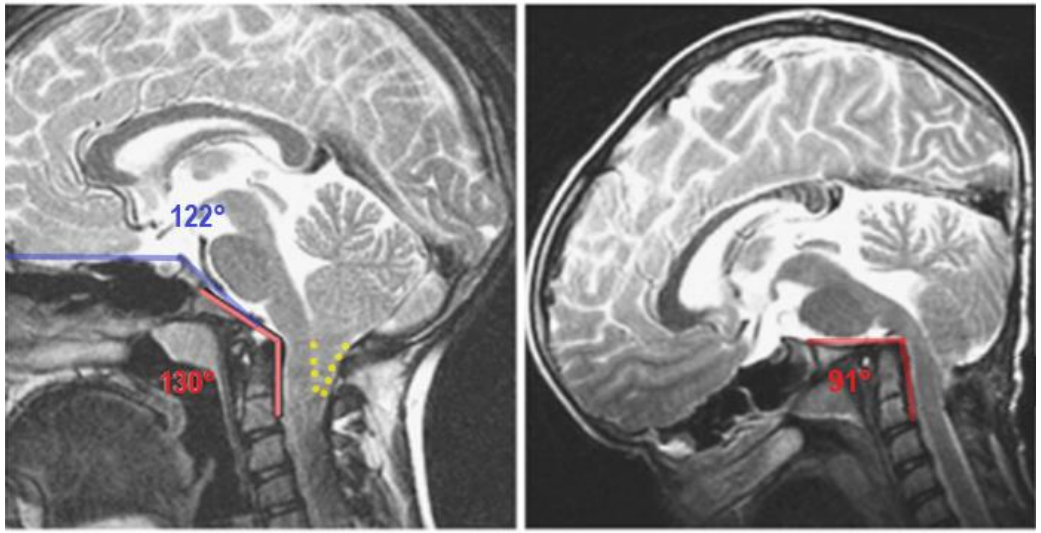




OAAJI instability, ISOL space occupying lesion, LPS shunt

Background – cervicomedullary syndrome

radiographic findings



- normal cranial base / clivus anatomy (NTB angle normal)
- craniocervical kyphosis (decrease in CXA), usually retroflexed odontoid (pBC2 ≥9mm)
- secondary tonsillar ectopia

diagnosis

- CXA <125-135°, NTB angle normal <130°, pBC2 ≥9mm
- pivoting of odontoid
- 4-6 weeks trial of immobilzation
- work-up bz connetive tissue expert

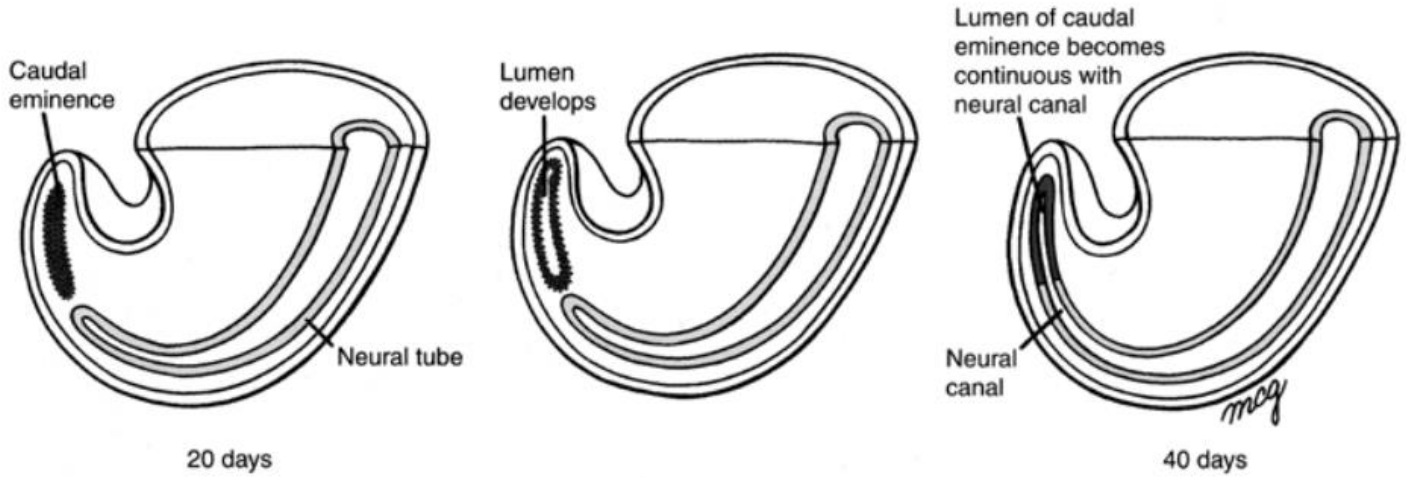
CT oder MRI in flexion/exntension, sitzend
Bei Tonsillenektopie: primär vs. sekundär (Volumetrie/Morphologie hintere Schädelgrube), Syringomyelie, Ausflusstrakt 4. Ventrikel

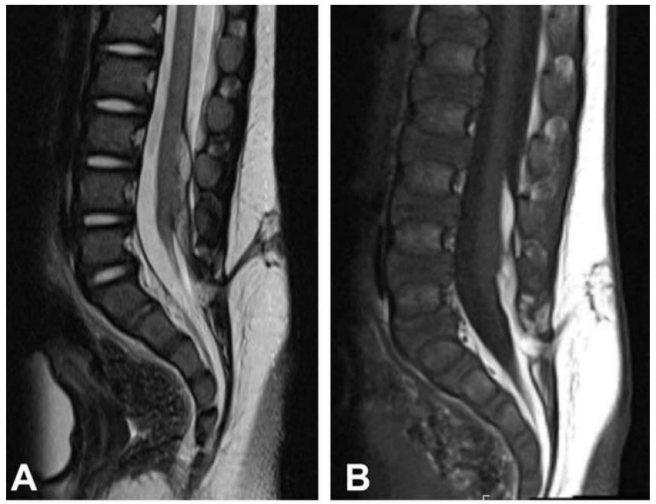
- Ravindra VM, Onwuzulike K, Heller RS, Quigley R, Smith J, Dailey AT, Brockmeyer DL. Chiari-related scoliosis: a single-center experience with long-term radiographic follow-up and relationship to deformity correction. J Neurosurg Pediatr. 2018 Feb;21(2):185-189. doi: 10.3171/2017.8.PEDS17318. Epub 2017 Nov 24. PMID: 29171800.

TABLE 1. Univariate analysis comparing patients who needed delayed thoracolumbar fusion and those who did not

Variable	Total Cohort	Delayed TL Fusion (11 pts)	No Delayed TL Fusion (12 pts)	p Value
Mos of follow-up	63.2 ± 55.6	88.3 ± 15.4	40.3 ± 14.7	**0.035**
Female sex	9/23 (39%)	6/11 (55%)	3/12 (25%)	0.214
Age at SODD (mos)	97.3 ± 55.3	91.6 ± 17.0	102.5 ± 16.2	0.65
No. of pts w/ CM 1.5 (%)	15/23 (65)	7/11 (64)	8/12 (67)	0.88
Tonsillar descent (mm)	9.02 ± 5.77	8.5 ± 1.8	9.5 ± 1.7	0.66
pBC2 (mm)	8.69 ± 2.48	9.56 ± 0.71	7.88 ± 0.69	0.11
No. of pts w/ pBC2 >9 mm (%)	10/23 (43)	7/11 (64)	3/12 (25)	**0.06**
CXA	139 ± 3.60	131.5 ± 4.8	146.5 ± 4.6	**0.034**
No. of pts w/ CXA >130° (%)	17/23 (74)	7/11 (64)	10/12 (83)	0.28
No. of syrinx levels	12.6 ± 6.3	11.5 ± 1.9	13.6 ± 1.8	0.43
No. of pts w/ holocord syrinx (%)	5/23 (22)	3/11 (27)	2/12 (17)	0.53
Initial Cobb angle (°)	29.6 ± 13.3	35.1 ± 3.6	22.8 ± 4.0	**0.035**
No. of pts w/ atypical curve (%)	13/23 (57)	7/11 (64)	6/12 (50)	0.89
No. of pts w/ levoscoliosis (%)	8/23 (35)	5/11 (45)	3/12 (25)	0.83
Time to fusion procedure (mos)	—	88.3 ± 15.4	—	—
No. of pts w/ chromosomal abnormality (%)	7/23 (30)	5/11 (45)	2/12 (17)	0.13

Hertzler DA 2nd, DePowell JJ, Stevenson CB, Mangano FT. Tethered cord syndrome: a review of the literature from embryology to adult presentation. Neurosurg Focus. 2010 Jul;29(1):E1. doi: 10.3171/2010.3.FOCUS1079. PMID: 20593997.

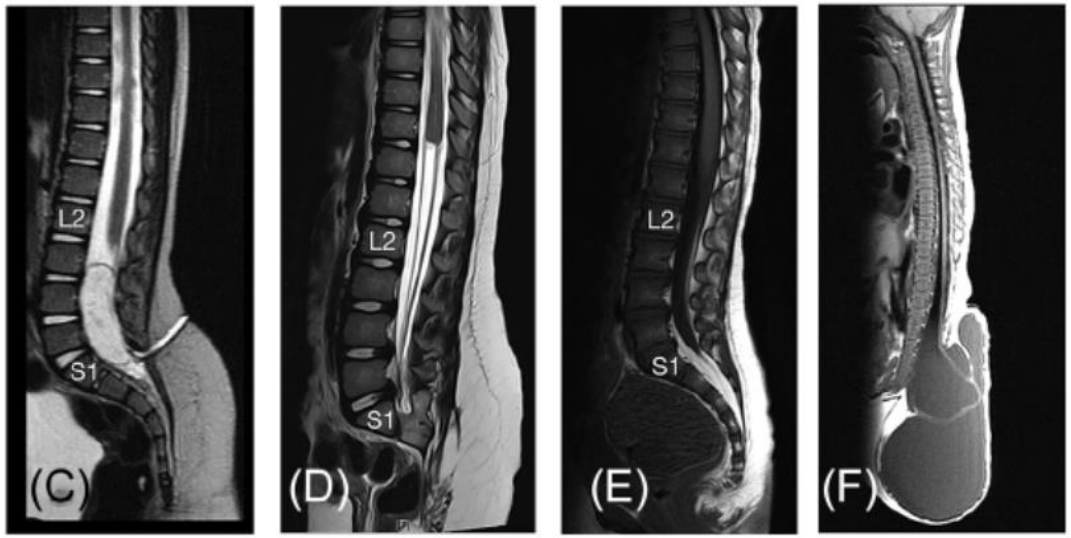




Tuite GF, Thompson DNP, Austin PF, Bauer SB. Evaluation and management of tethered cord syndrome in occult spinal dysraphism: Recommendations from the international children's continence society. Neurourol Urodyn. 2018 Mar;37(3):890-903. doi: 10.1002/nau.23382. Epub 2017 Aug 9. PMID: 28792087.

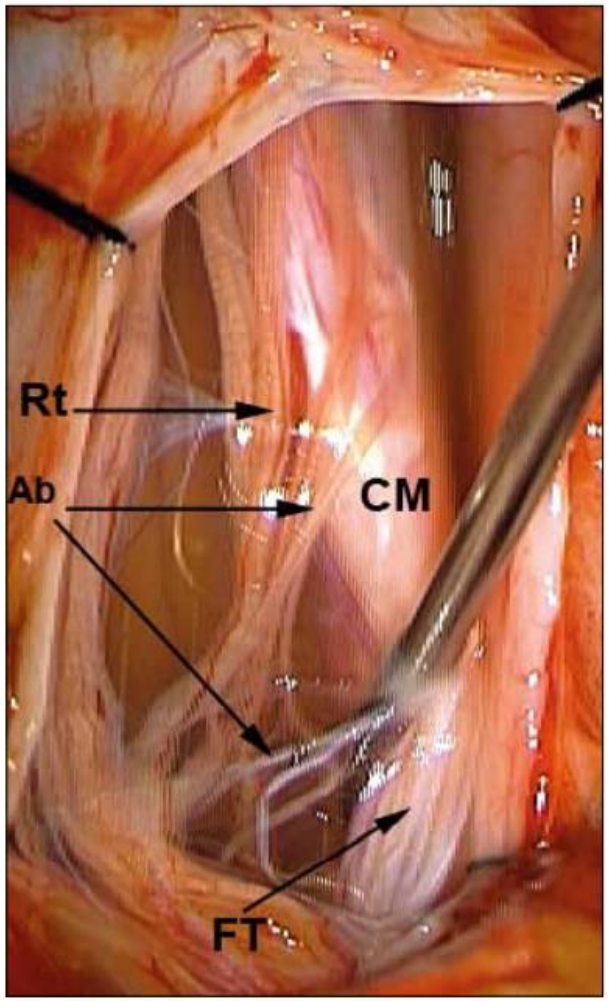
Klassisches tethered cord

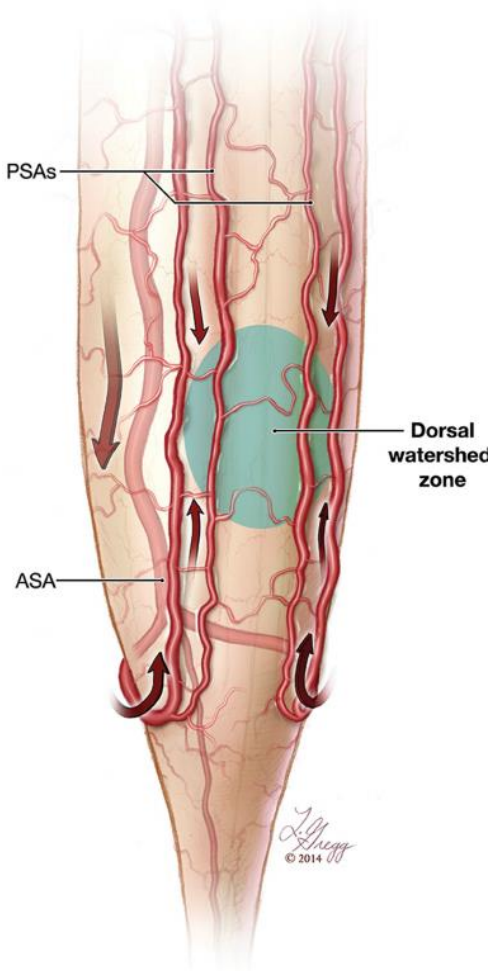
Normaler Erwachsener
Conus L2/3
Filum dicke (2mm)?
Fett



	Developmental stage				
		Primary neurulation			
			Dysjunction		
	Gastrulation	Neural tube closure	Premature	Incomplete	Secondary neurulation
Dysraphism diagnosis	Split cord malformations	Myelomeningocele	Lipoma (dorsal)	Dermal sinus track	Thickened filum
	Neurenteric cysts			Limited dorsal myeloschisis	Lipoma (caudal, transitional and chaotic)
					Terminal myelocystocele

Gailloud P, Gregg L, Galan P, Becker D, Pardo C. Periconal arterial anastomotic circle and posterior lumbosacral watershed zone of the spinal cord. J Neurointerv Surg. 2015 Nov;7(11):848-53. doi: 10.1136/neurintsurg-2014-011408. Epub 2014 Oct 3. PMID: 25280570.





Klassische Symptome:
LumboSAKRALGIEN
Nicht dermatomale BeineSz (brennend, Muskelkaterartig, ähnlich neuropathisch)
Beine schwer, steif

Neuro Untersuchung
Kinder:
Erwachsene: Hyperreflexie, Spasti

Neurogene Blasenfunktionsstörung

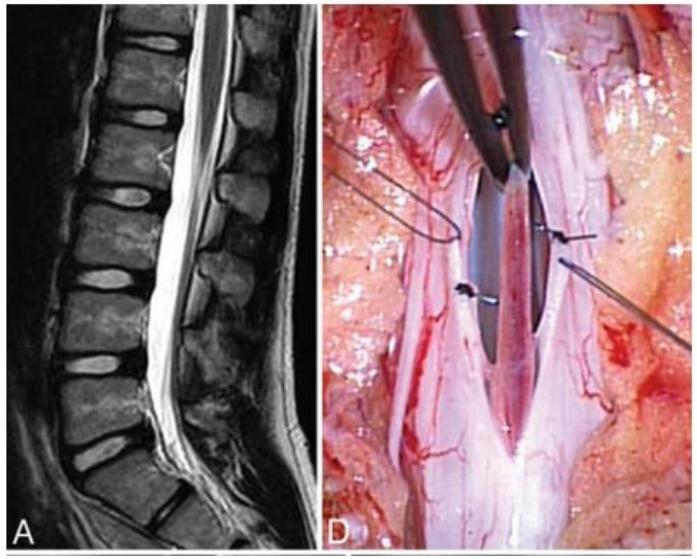
Orthopädische Fuss (oder Skoli)

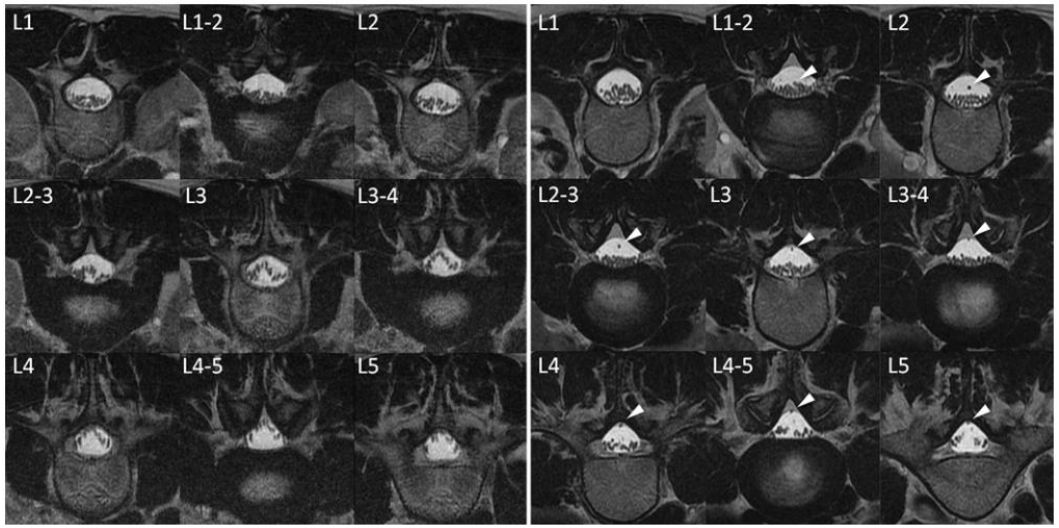
Gailloud P, Gregg L, Galan P, Becker D, Pardo C. Periconal arterial anastomotic circle and posterior lumbosacral watershed zone of the spinal cord. J Neurointerv Surg. 2015 Nov;7(11):848-53. doi: 10.1136/neurintsurg-2014-011408. Epub 2014 Oct 3. PMID: 25280570.

Diagnose 3 von 4 Kategorien

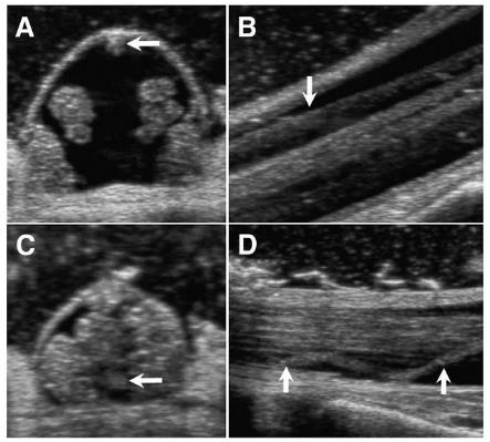
Neurologie/Anamnese
Orthopädische Manifestationen
Urologische Manifestiationen
Hauterscheinungen

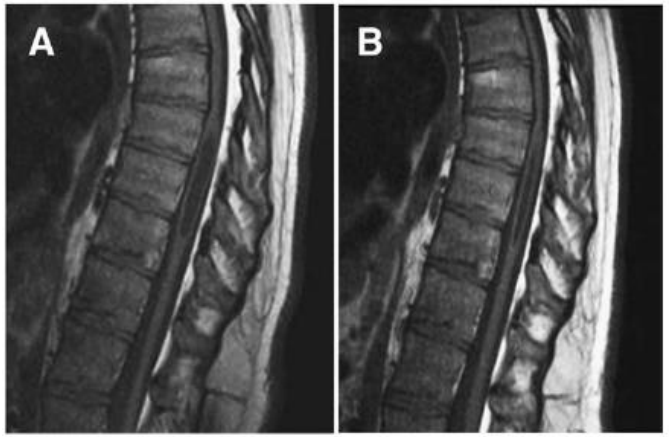
(bei indirekten radiologischen Zeichen
ist Diagnose auch mit weniger als 3
Kategorien möglich)
Syrinx
Filum dick oder Filum getetherd
posterior

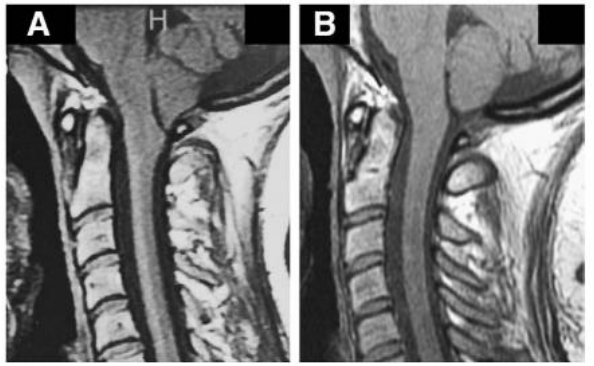




- Nakanishi K, Tanaka N, Kamei N, Nakamae T, Izumi B, Ohta R, Fujioka Y, Ochi M. Use of prone position magnetic resonance imaging for detecting the terminal filum in patients with occult tethered cord syndrome. J Neurosurg Spine. 2013 Jan;18(1):76-84. doi: 10.3171/2012.10.SPINE12321. Epub 2012 Nov 9. PMID: 23140126.







Milhorat TH, Bolognese PA, Nishikawa M, Francomano CA, McDonnell NB, Roonprapunt C, Kula RW. Association of Chiari malformation type I and tethered cord syndrome: preliminary results of sectioning filum terminale. Surg Neurol. 2009 Jul;72(1):20-35. doi: 10.1016/j.surneu.2009.03.008. Erratum in: Surg Neurol. 2009 Nov;72(5):556. PMID: 19559924; PMCID: PMC5999045.

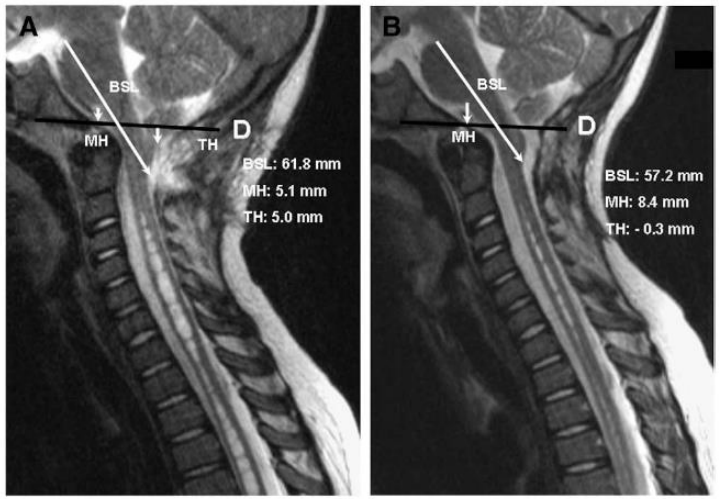


Fig. 7. Morphometric measurements of CCJ before and after SFT in 8-year-old female with CM-I/TCS and slowly progressive syringomyelia 3 years after Chiari decompression surgery had failed to reduce syrinx size. Reconstructed CT scans of head revealed complete reossification of suboccipital craniectomy. A: Midsagittal T-2 weighted MR image before SFT showing elongation of brain stem (BSL = 61.8 mm), downward displacement of medulla (MH = 5.1 mm), herniation of cerebellar tonsils (TH = 5.7 mm), and persistent cervicothoracic syringomyelia. B: Postoperative scan 5 weeks after SFT showing reduction of BSL (57.2 mm), ascent of medulla (MH = 7.4 mm), ascent of cerebellar tonsils (TH = 0), expansion of upper cervical subarachnoid spaces, and reduction in size of syringomyelia.

Syrinx: Chiari, TCS, Tumor (cave Hemangioblastoma), unspecific (compression, tethering)

Milhorat TH, Bolognese PA, Nishikawa M, Francomano CA, McDonnell NB, Roonprapunt C, Kula RW. Association of Chiari malformation type I and tethered cord syndrome: preliminary results of sectioning filum terminale. Surg Neurol. 2009 Jul;72(1):20-35. doi: 10.1016/j.surneu.2009.03.008. Erratum in: Surg Neurol. 2009 Nov;72(5):556. PMID: 19559924; PMCID: PMC5999045.

Metcalfe PD, Luerssen TG, King SJ, Kaefer M, Meldrum KK, Cain MP, Rink RC, Casale AJ. Treatment of the occult tethered spinal cord for neuropathic bladder: results of sectioning the filum terminale. J Urol. 2006 Oct;176(4 Pt 2):1826-9; discussion 1830. doi: 10.1016/j.juro.2006.04.090. PMID: 16945660.

TABLE 1. *Significant pathological findings in our cohort despite maximal medical therapy**

Symptom	% Prevalence
Daytime urinary incontinence	83
Nocturnal enuresis	78
Recurrent urinary tract infection	36
Constipation	69
Encopresis	50

* Including maximal anticholinergic therapy, an aggressive bowel program and biofeedback training.

TABLE 2. *Symptoms elicited by neurosurgeon that were not detected by urologist*

Symptom	No. Pts
Leg or foot pain	7
Back pain	6
Increased pain with exercise	6
Decreased perineal sensation	3
Lower extremity clumsiness	3
Total	25

There were 20 patients with these 25 extra-urinary symptoms.

TABLE 5. *Patients who reported any degree of improvement in urinary symptoms*

Symptom	% Overall Improvement	% Cure	
		At 3 Mos	Within 1 Yr
Urinary symptoms	72		
Daytime incontinence	68	26	40
Urgency	64	45	73
Frequency	40	40	50
Nocturnal enuresis	36	21	40
Constipation	92	32	40
Encopresis	83	50	61

Patient or caregiver subjectively defined improvement, urgency, frequency and constipation with incontinence results based on the presence or absence as reported by the caregiver.

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TABLE 1. *Significant pathological findings in our cohort despite maximal medical therapy**

Symptom	% Prevalence
Daytime urinary incontinence	83
Nocturnal enuresis	78
Recurrent urinary tract infection	36
Constipation	69
Encopresis	50

* Including maximal anticholinergic therapy, an aggressive bowel program and biofeedback training.

TABLE 2. *Symptoms elicited by neurosurgeon that were not detected by urologist*

Symptom	No. Pts
Leg or foot pain	7
Back pain	6
Increased pain with exercise	6
Decreased perineal sensation	3
Lower extremity clumsiness	3
Total	25

There were 20 patients with these 25 extra-urinary symptoms.

TABLE 5. *Patients who reported any degree of improvement in urinary symptoms*

Symptom	% Overall Improvement	% Cure	
		At 3 Mos	Within 1 Yr
Urinary symptoms	72		
Daytime incontinence	68	26	40
Urgency	64	45	73
Frequency	40	40	50
Nocturnal enuresis	36	21	40
Constipation	92	32	40
Encopresis	83	50	61

Patient or caregiver subjectively defined improvement, urgency, frequency and constipation with incontinence results based on the presence or absence as reported by the caregiver.

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CASE REPORT

Occult tethered cord syndrome: a reversible cause of paraparesis not to be missed

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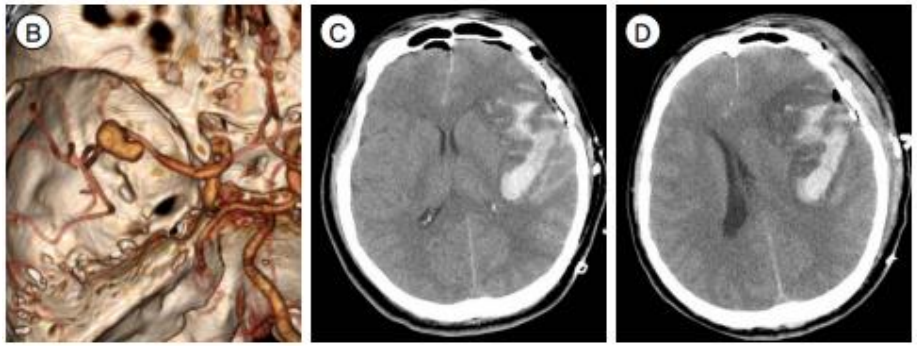
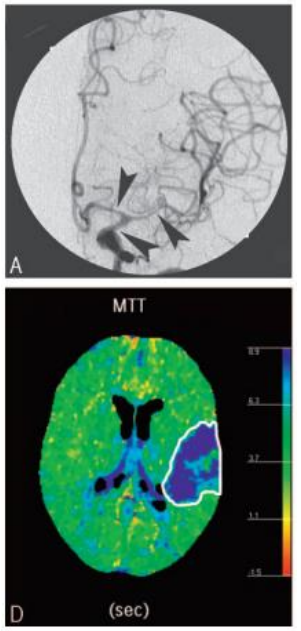
Abstract

A 15-year-old female former gymnast with a history of pectus excavatum was reviewed due to unexplained paraparesis and urinary incontinence since age 10. Symptoms were commenced with intolerable upper back pain and development of a soft mass at the sacrum that remitted spontaneously. Brain and whole spine MRI imaging and blood and CSF testing were normal. The combination of skeletal, neurological, and bladder symptoms with normal lumbar MRI and abnormal urodynamic and neurophysiological studies led to the clinical suspicion of occult tethered cord syndrome (oTCS). Surgical cord "untethering" was performed leading to remarkable postoperative clinical improvement. oTCS is a recently defined functional disorder of the spinal cord due to fixation (tethering) of the conus medullaris by inelastic elements that may lead to severe neurological impairment. High clinical suspicion is required as oTCS is a treatable spinal cord disorder.

¾ Diagnosepunkte
Ggf. MRI LWS in Bauchlage
Assozierte Syrinx, Skoliose, hintere schädelgrube





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