

Foramen magnum stenosis (FMS): neuroradiological aspects before and after cervical decompression in paediatric patients with achondroplasia (ACH)

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Background and Aim

The identification of anamnestic, clinical and instrumental data indicative of pathological FMS plays a pivotal role in the prevention of ACH complications.

Aim: identify key cranio-cervical junction(CCJ)neuroradiological features for the surgical choice and for the neuroradiological decompression outcome.

Subjects and Methods

From a **total of 191 patients**, we selected **24 subjects with ACH (age:<4years)**, who performed a first brain MRI and/or CT.

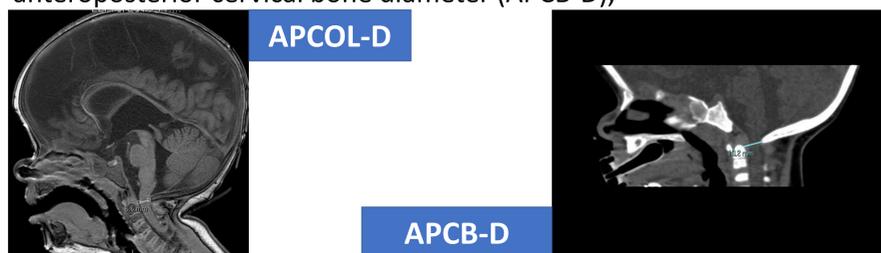
Patients were divided into 2 groups:

- 1.surgically treated patients (STP=15/24)
- 2.non-surgically treated patients (NSTP=9/24).

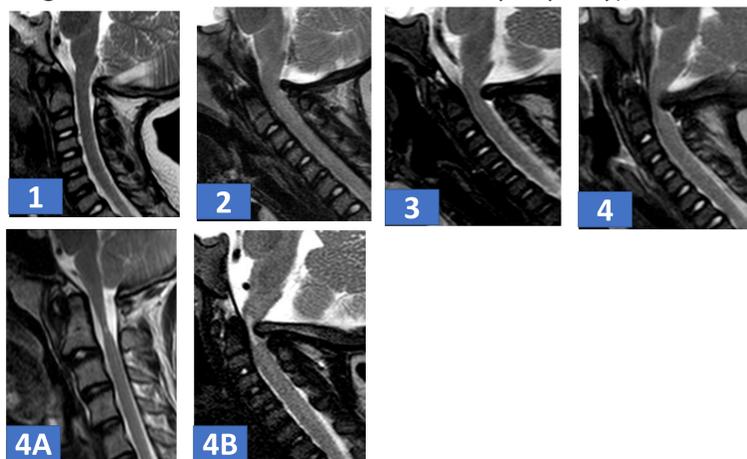
The data were compared with a control group (CG) of 24 children of the same age and with a group of ACH patients surgically treated at an age of more than 4 years (ACHPST>4AA, 5/191).

MRI evaluation

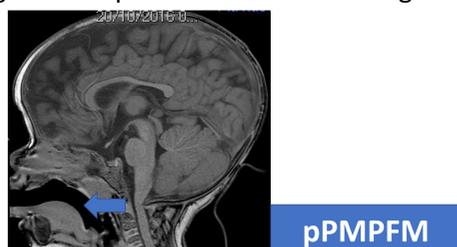
- antero-posterior cervical osteo-ligamentous diameter (APCOL-D)
- anteroposterior cervical bone diameter (APCB-D),



- degree of cervical stenosis (grade 0, 1, 2, 3, defined respectively on the basis of the increase in stenosis grade 4A and 4 B defined according to the degree of stenosis in association with myelopathy),



- posterior margin of the prominent foramen magnum (prominence PMPFM),



- posterior arc of prominent C1,
- hypertrophy of soft tissues, occipital bone spur,
- orientation of the posterior edge of the foramen magnum,

Results

33.3% of subjects who performed the first MRI in the first 6 months of life have myelopathy (stenosis 4A and 4B)

-All STP have cervical stenosis of grade>2 while the NSTP have degrees< 2.

-Grade 1 is equally represented in STP and NSTP.

-APCOL-D is significantly lower in:
STPvsNSTP (P<0,0001)
STPvsCG ((P<0,0001)
NSTPvsCG (P<0,0001) with an OR=3.95 (P=0.02).

-The APCB-D is significantly lower in:
the STPvsCG (P<0.0001)
the NSTPvsCG (P<0.001)
there is no significant difference between STPvsNSTP.

-Prominence PMPFM is significantly associated with surgery (p=0.003).
No other qualitative parameters are significantly associated.

-In STP there is a significant increase of APCOL-D and APCB-D (p = 0.0001).

Conclusion

- Brain MRI is crucial in the preventive diagnosis of complications (screening role).**
- The importance of performing MRI in the first 6 months of life has been highlighted.**
- The most important radiological parameters for surgical choice are:**

prominence PMPFM

the APCOL-D
(values<7.6mm determine a risk of surgical therapy 4 times higher)

degrees of stenosis>2.

- STP have a very good radiologic decompression outcome.**

The data of this pilot study will be correlated with multidisciplinary approach, useful in particular in the evaluation of grade1 stenosis (still grey area regarding surgical choice).

References

- Developmental Medicine & Child Neurology 2017, 59: 192–198J
Neurosurg Pediatrics 2:136–138, 2008
American Journal of Medical Genetics, 31 August 2015: 42-51
Orphanet J Rare Dis. 2019 Jan 3;14(1):1.