

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

Allegri AEM¹, Di Iorgi N¹, Severino M², Michelis B⁴, Patti G¹, Siri G¹, Fava D¹, Calandrino A¹, Piatelli G³, Maghnie M¹.
The 'Achondroplasia Multidisciplinary Gaslini's Group' (AMGG)

¹.Paediatric Clinic, Gaslini Children's Hospital Genova, Italia; ².Neuroradiology Gaslini Children's Hospital Genova, Italia; ³.Neurosurgery Gaslini Children's Hospital Genova, Italia, ⁴.Paediatric Orthopedics Unit Gaslini Children's Hospital Genova

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.

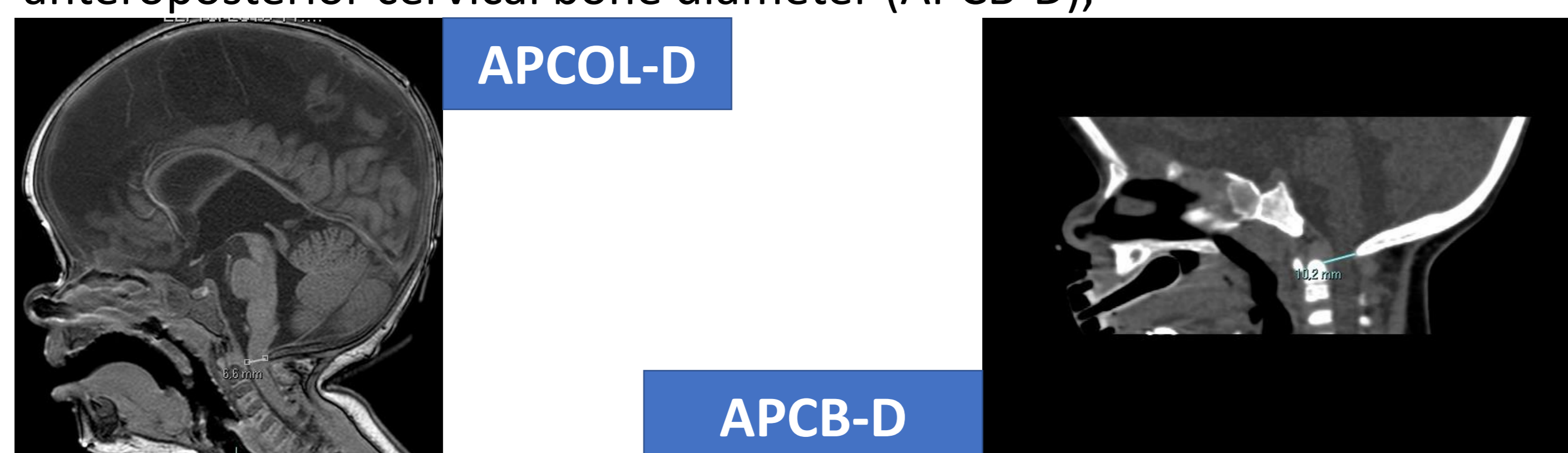
Patients were divided into 2 groups:

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

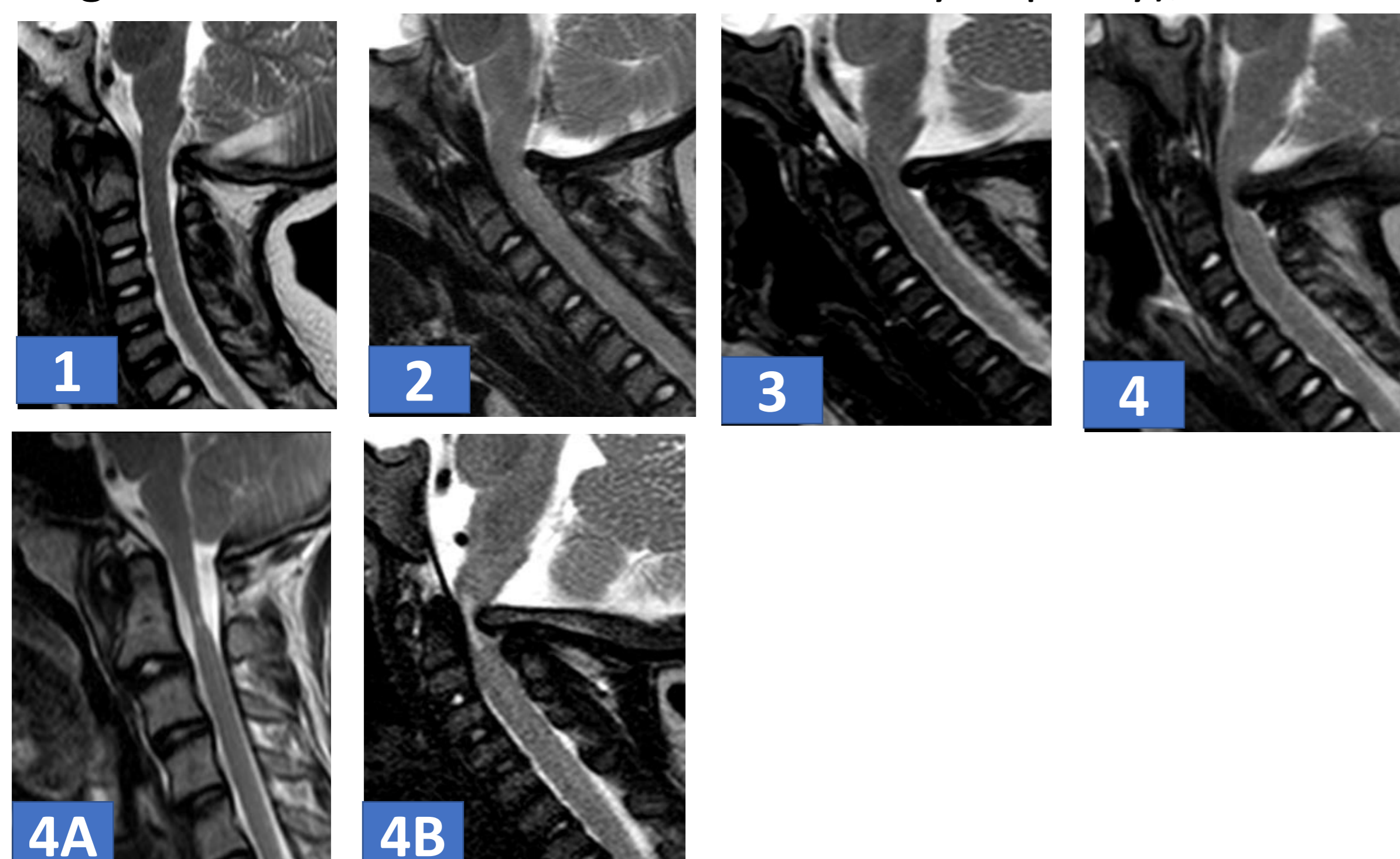
The data were compared with a control group (CG) of 24 children of the same age and with a group of ACH patients surgically treated at an age of more than 4 years (ACHPST>4AA, 5/191).

MRI evaluation

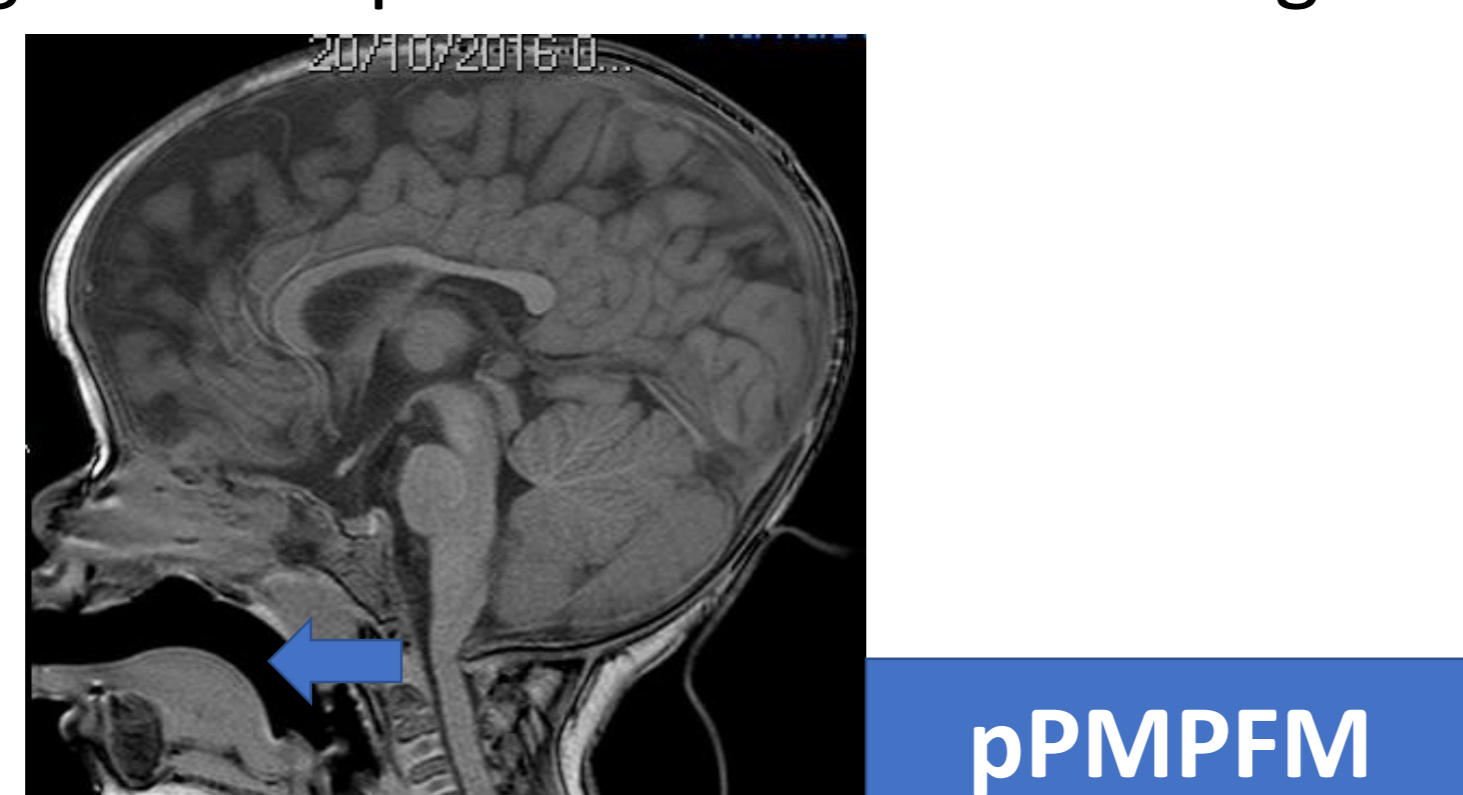
- antero-posterior cervical osteo-ligamentous diameter (APCOL-D)
- anteroposterior cervical bone diameter (APCB-D),



- degree of cervical stenosis (grade 0, 1, 2, 3, defined respectively on the basis of the increase in stenosis grade 4A and 4B 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.