Oral presentation Open Access **Reliability of the MRI diagnosis of the Chiari type2 malformation** Reinier Mullaart^{*1}, Niels Geerdink¹ and Ton van der Vliet²

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Background

Every clinician familiar with spina bifida knows what Chiari type2 malformation (C2M) is about. However, as soon as it concerns a single patient, the discussion starts whether or not it is justified to make the diagnosis in that particular case. This complicates the insight in the epidemiology and pathogenesis, complicates the indication for (fetal) surgery and complicates inferences on the effect of treatment. This was reason for us to investigate the reliability of the diagnosis of C2M.

Materials and methods

We investigated the reliability of the diagnosis of C2M, by testing the inter observer correlation of 49 MRI criteria which in the literature are considered specific for C2M. With this aim two investigators reviewed MRIs of presumed C2M cases and healthy controls (n = 10). The images were blinded for demographic and clinical information, subsequently arranged by projection (sagittal, axial, coronal) and finally by individual. As a consequence, the connection between the different projections per individual was taken away. Yes/no criteria with more than 80% agreement were considered sufficiently reliable. For quantitative criteria a Spearman rho correlation coefficient of 0.9 was taken as boundary.

Results

Seven out of 29 yes/no criteria and seven out of twenty quantitative criteria fulfilled the requirements of reliability (\geq 80% agreement or Spearman rho \geq 0.9). These criteria were: downward herniation of the vermis; herniation

around the brain stem; narrowed 4th ventricle; mesencephalic beaking; flattening of the pons; kinking of the medulla; and qualitative measures as position of tonsil, vermis and 4th ventricle; tentorium length; pons thickness; and cerebellar width. The majority of these criteria were obtained from sagittal images (11/29 sagittal criteria); a few from the other images (1/7 coronal criteria; 2/ 14 axial criteria). Thirty-five criteria did not fulfil the requirements of reliability; some were virtually immeasurable.

Conclusion

A considerable number of criteria that are by tradition considered specific for C2M seem quite unreliable. Against this, a considerable set of criteria, in particular those in the sagittal projection, remains helpful for the diagnosis. Analysis of a larger sample is in progress with the aim to further substantiate the present findings and to make inferences on the specificity of the criteria for C2M.