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The association of Chiari malformation type 1 and closure of sagittal suture

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Introduction: The association between Chiari Malformation type 1 (CM1) and non-syndromic sagittal synostosis is estimated between 1.4% and 5.8% However, the real incidence of this association is still debated.

Methods: To study such association, several analyses were performed:
1 - a retrospective review of 912 consecutive surgically corrected scaphocephalies
2 - a review of 38 consecutive operated on CM1 patients
3 – a review of 100 control CTs (head trauma with normal CT) .
4 - a review of 11 non operated scaphocephalies (parents refused).
The pre-operative CT were analysed in all operated on children and last follow-up images for the non operated scaphocephalies. The cervical-medullary MRIs were examined in the CM1 patients to define: the degree of tonsillar herniation and the presence of hydrocephalus and/or syringomielia.

Results: Three out of 912 (0,3%) children with scaphocephaly had a CM1 at diagnosis (mean age 3 months). None of the non operated scaphocephaly (mean age 9.2 years) had a CM1.
Among the 38 symptomatic CM1 patients (mean age 7.7 years). , 8 (21%) showed a closed sagittal suture (mean age 7.5 years). Bone associated syndromes were more common in this subgroup (37.5% in CM1 with fused sagittal suture compared to 17.4% in CM1 with open sagittal suture). No other statistical differences were found between the two subgroups.
In the control group, (mean age 9.2 years) the sagittal suture appeared fused in 4%.

Discussion: CM1 associated to the fusion of the sagittal suture seems to be a different nosological entity that a “classical” scaphocephaly (with an early fusion diagnosed in infancy) and that isolated symptomatic CM1. This distinction highlights the importance of complete workout in patient presenting with CM1 and might have an impact in the surgical strategy.