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Pathogenesis of cerebral malformations in perinatal spina bifida aperta

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Background

Spina bifida aperta (SBA) is associated with cerebral morbidity, such as hydrocephalus, Chiari II malformation and cortical dysplasia. Insight in the pathogenesis of these malformations is incomplete. In fetal SBA, such information may help to improve pre- and early postnatal treatment strategies. In perinatal SBA, we investigated the time of initiation of concurrent cerebral malformations.

Materials and methods

In 7 SBA fetuses and 1 neonate [16–40 (median 28) weeks gestational age (g.a.)], we cross-sectionally investigated the histology of the aqueduct [n = 5], cerebral convexity and parenchyma [n = 8] by haematoxylin-eosin and nestin staining. The meningomyelocele was located at cervical [n = 1], thoracic [n = 3] and lumbar [n = 4] spinal level. Cerebral histology was intra-individually associated with fetal ultrasound parameters (ventricular size, head circumference and Chiari II malformation). The mean and median duration between fetal ultrasound and histological assessment were both 4 days.

Results

In SBA fetuses of all gestational ages, histological malformations at the aqueduct (hemosiderophages/gliosis [5/5] and forking/slit like deformities [5/5]) were present. In the two youngest fetuses (16 and 21 weeks g.a.), we observed peri-aqueductal ependymal denudation, progenitor cell loss and heterotopia. From the 2nd half of pregnancy onwards, Chiari II malformation concurred with ventriculomegaly [4/6] and successively, with macrocephaly from 37 weeks g.a. onwards [3/3]. In absence of arachnoidal fibrosis, delivery-related haemorrhages were present in all fetuses (at the fossa posterior and/or cerebrum in 6/7 and 5/7 fetuses, respectively). In the only patient that succumbed during the first week after birth (39 weeks g.a.), raised intracranial pressure concurred with arachnoidal fibrosis at the convexity.

Conclusion

In fetal SBA, the earliest peri-aqueductal alterations precede the development of hydrocephalus. During the 2nd half of pregnancy, ventriculomegaly appeared unrelated to CSF malabsorption. After birth, however, CSF malabsorption may increasingly contribute to the development of high-pressure hydrocephalus. These data may implicate that peri-aqueductal ependymal denudation and progenitor cell loss occur by a mechanism independent of high-pressure hydrocephalus or ventricular distention.