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Perforation of the Cavum Septi Pellucidi in Open Spina Bifida: Association with Need for Hydrocephalus Treatment in the First Year of Life

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Objective

To measure the association of detecting a perforated cavum septi pellucidi (CSP) seen on brain imaging of fetuses referred to a large Fetal Center for open neural tube defect (ONTD) repair at the time of referral and six weeks postoperatively, with the eventual need for hydrocephalus treatment during the first year of life.

Methods

Retrospective cohort study of 110 patients who underwent laparotomy-assisted fetoscopic ONTD repair between 2014 and 2021 at a single center. Eligibility criteria for surgery were based on MOMS trial criteria although maternal BMI up to 40 kg/m2 was allowed. Fetal brain imaging was assessed with ultrasound and MRI at initial referral and six weeks postoperatively. Retrospective review to assess CSP integrity was performed using stored US and MRI scans. Information on the need for hydrocephalus treatment by 12 months was obtained by medical record review. Parametric tests and non-parametric tests were used as appropriate to compare outcomes between pCSP vs intact CSP cases as assessed by ultrasound at referral. Logistic regression analyses were performed to assess predictive values for the need for a hydrocephalus treatment.

Results

A total of 20.6% and 21.8% cases were identified with a preoperative pCSP by US and MRI, respectively, and 26.6% and 24.2% on post-operative US and MRI, respectively. Ventricular size increased after surgery. Assessment of 107 cases with complete CSP evaluation by US at referral were included for comparison: 22 with pCSP and 85 with intact CSP. The pCSP group presented with larger ventricles (14.32 +3.45 mm vs. 10.37^{+2}.37 mm); p<0.01) and more severe ventriculomegaly (40.9% vs. 5.9%; p<0.01). The same associations were observed at 6 weeks postoperatively [ventricular size: 21 mm (13-43.5 mm) vs. 14.25 mm (7-29 mm); p<0.01], and [severe ventriculomegaly: 95% vs. 46.8%; p<0.01]. Cases with pCSP had a lower rate of hindbrain herniation (HBH) reversal postoperatively (65% vs. 88.6%; p=0.01) [preoperatively: 11 (5.89-21.45) mm vs. postoperatively: 16 (7-43.5) mm; p<0.01] as well as the proportion of cases with severe ventriculomegaly [12.7% vs 57.8%; p<0.01]. Cases with pCSP required treatment for hydrocephalus more frequently than those without that feature [17/19 (89.5%) vs. 17/75 (22.7%); p<0.01]. The strongest predictor for this outcome was the lack of HBH reversal (OR 36.2, 95% CI 5.96-219.12; p<0.01) followed by pCSP at referral (OR 23.4, 95% CI 5.42-100.98, p<0.01) and by pCSP at 6 weeks postop (OR 19.48, 95% CI 5.68-66.68, p<0.01).

Conclusion

The detection of a pCSP in fetuses with ONTD can reliably identify those cases at highest risk for needing hydrocephalus treatment during the first year of life. The evaluation of this brain structure can greatly improve our counseling to families considering fetal surgery for ONTD, in order to set expectations about postnatal outcome.