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Growing bone defect in an infant with a rare combination of a foramen parietale per magna and an atretic cephalocele

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Introduction: We present a case with an atretic cephalocele and large persistent parietal foramina that increased in size over time and were revealed during surgery. Large parietal foramina are a rare clinical entity with a prevalence ranging from 1:15,000 to 1:25,000. The skull defect is usually identified on physical examination and confirmed radiographically.

Methods: The female patient was presented to our department at 2 months of age. She was born mature. After birth a soft swelling was noted parietooccipital, over time the swelling was documented to grow in size. An MRI showed an occipital cephalocele surrounded by cerebrospinal fluid. The patient had no other neurological symptoms. During surgery the subdural collection of cerebrospinal fluid was identified as enlarged parietal foramina measuring 3x2 and 2x2.5 cm. The brain tissue was elevated above bone level without sufficient dura cover. Surgical treatment involved covering the brain tissue with an artificial dura substitute (DuraGen Plus™) and performing cranioplasty using autologous bone fragments.

Results: The cosmetic result was satisfying. There have been no problems with wound healing and no new neurological deficits occurred. Long-term follow-up will be performed.

Conclusions: Cephaloceles have rarely been associated with enlarged parietal foramina. Only one other case was published. Developmentally enlarged parietal foramina arise as ossification defects involving the parietal bone. If the defect occurs bilaterally it is subsequently cleaved into two foramina by parasagittal islands of ossification. Typically the cleft decreases in size during development. Also to our knowledge this is the only case in which the parietal foramina increased in size at such an early age. We hypothesize this is due to the fact that no dura was covering the pulsating brain underneath the bone defect, comparable with growing skull fractures resulting in the need of surgical closure.