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Encephalocele – Experience in a series of 42 cases

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Introduction: Encephalocele is a rare congenital anomaly of the central nervous system, accompanied by defects in the skull and dura with herniation of brain tissue, meninges and cerebrospinal fluid. The aim of this study was to perform a descriptive analysis of a series of cases of children with encephalocele.

Methods: Retrospective, descriptive, multicenter study was conducted between 1995-2009 in a Beneficencia Portuguesa Hospital and Hospital Santa Marcelina - Itaquera, São Paulo - SP. A series of 42 surgeries to repair encephalocele is described. The children studied were operated aged between 12 hours and 6 years old. A descriptive analysis of clinical aspects presented by the cases studied was performed.

Results: Forty patients were diagnosed with encephalocele, with the highest prevalence for females (22 women). Regarding the anatomical location, 30% were anterior/ fronto-facial (12 patients) and 70% posterior / ocipital (28 patients). In the anterior localization. 37 in the vertex (92.5% and 3 frontoethmoidal / sincipital (7.5%). Eleven patients (27.5%) were associated with other pathologies: hydrocephalus (4 cases, 10%) , amniotic band (5 cases, 12.5%), cardiac malformation (1 case, 2.5%), agenesis cutis (1 case, 2.5%) and microcephaly (1 case, 2.5%). Two patients underwent an adicional surgical procedure to treat hydrocephalus, wound dehiscence.

Conclusion: Posterior encephalocele was more frequent in this series. Apoximately 27.5% of the cases presented other congenital anomalies. The most common neurological malformation associated with encephalocele was hydrocephalus.