Risk factors for hydrocephalus and neurological deficit in children born with an encephalocele

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Abstract

OBJECT—There is a known association of hydrocephalus with encephaloceles. Risk factors for hydrocephalus and neurological deficit were ascertained in a series of patients born with an encephalocele.

METHODS—A retrospective analysis was undertaken of patients treated for encephaloceles at Children's Hospital Los Angeles between 1994 and 2012. The following factors were evaluated for their prognostic value: age at presentation, sex, location of encephalocele, size, contents, microcephaly, presence of hydrocephalus, CSF leak, associated cranial anomalies, and neurological outcome.

RESULTS—Seventy children were identified, including 38 girls and 32 boys. The median age at presentation was 2 months. The mean follow-up duration was 3.7 years. Encephalocele location was classified as anterior (n = 14) or posterior (n = 56) to the coronal suture. The average maximum encephalocele diameter was 4 cm (range 0.5–23 cm). Forty-seven encephaloceles contained neural tissue. Eight infants presented at birth with CSF leaking from the encephalocele, with 1 being infected. Six patients presented with hydrocephalus, while 11 developed progressive hydrocephalus postoperatively. On univariate analysis, the presence of neural tissue, cranial anomalies, encephalocele size of at least 2 cm, seizure disorder, and microcephaly were each positively associated with hydrocephalus. On multivariate logistic regression modeling, the single prognostic factor for hydrocephalus of borderline statistical significance was the presence of neural tissue (odds ratio [OR] = 5.8, 95% confidence interval [CI] = 0.8–74.0). Fourteen patients had severe developmental delay, 28 had mild/moderate delay, and 28 were neurologically normal. On univariate analysis, the presence of cranial anomalies, larger size of encephalocele, hydrocephalus, and microcephaly were positively associated with neurological deficit. In the

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multivariable model, the only statistically significant prognostic factor for neurological deficit was presence of hydrocephalus (OR 17.2, 95% CI 1.7–infinity).

CONCLUSIONS—In multivariate models, the presence of neural tissue was borderline significantly associated with hydrocephalus and the presence of hydrocephalus was significantly associated with neurological deficit. The location of the encephalocele did not have a statistically significant association with incidence of hydrocephalus or neurological deficit. In contrast to modestly good/fair neurological outcome in children with an encephalocele without hydrocephalus, the presence of hydrocephalus resulted in a far worse neurological outcome.

Keywords

congenital abnormality; encephalocele; hydrocephalus; neurological outcome

With an estimated global prevalence of 0.8–4 per 10,000 live births,[1,19,20,26–28,30,31] encephaloceles, along with anencephaly and myelomeningocele, are common forms of neural tube defects. Defined as cystic congenital malformations in which CNS structures, in communication with CSF pathways, herniate through a defect in the cranium,[25] encephaloceles are generally classified based on anatomical location. Infants with encephaloceles may often have associated malformations, reported to occur in 36%–60% of patients,[22,32] and include anomalies such as microcephaly, absence of the corpus callosum, optic nerve abnormalities, hypothalamic/pituitary dysfunction, fusion of thalami, schizencephaly, holoprosencephaly, arachnoid cyst, Dandy-Walker malformation, Walker-Warburg syndrome, Chiari malformation, craniosynostosis, hypertelorism, hemifacial microsomia, cleft lip/palate, Klippel-Feil malformation, myelomeningocele, and hydrosyringomyelia.[3,5,17,22,27,34]

The prognosis for a newborn with an encephalocele depends on a number of factors and includes the location, size, and contents of the encephalocele, along with microcephaly and associated anomalies.[4,16,27,34] A significant proportion of these children lack normal developmental milestones and may have neurological problems such as ataxia, visual impairment, mental and growth retardation, and seizures.[22] Previous studies report that prognosis for patients with occipital and parietal encephaloceles is directly related to the presence of neural tissue, hydrocephalus, microcephaly, and accompanying cerebral malformations.[6,10,14,20] Hydrocephalus carries a significant morbidity in these children, reported to occur in as many as 60%–90% of patients with posterior encephaloceles, in contrast to a much lower incidence of 10%–15% in anterior encephaloceles.[2,13,23] To date, the risk factors associated with hydrocephalus requiring CSF diversion in infants born with an encephalocele have not been fully investigated. In this study, we review our series of patients diagnosed with an encephalocele and analyze the risk factors for hydrocephalus and neurological deficit in these pediatric patients.

Methods

Study Population

We performed a retrospective review of all patients who presented to Children's Hospital Los Angeles with an encephalocele over an 18-year period (1994–2012). Patients included

in the study were those diagnosed with encephalocele and who underwent primary surgical repair by the Division of Neurosurgery at our institution. Patients who underwent primary surgical repair of their encephalocele at an outside hospital were excluded from the study as pertinent data were often unavailable. Atretic encephaloceles, considered to be noncystic—i.e., no CSF, nodular or flat, small midline, and posteriorly located behind the coronal suture—were also excluded. Medical records of eligible patients were reviewed by the primary author to extract the following variables: age at presentation, race/ethnicity, sex, microcephaly, seizure disorder, location of encephalocele, size, contents, presence of hydrocephalus, CSF leakage, associated cranial anomalies, and neurological outcome. Encephalocele location was classified as anterior or posterior based on its relation to the coronal suture. Contents in the encephalocele were initially defined by imaging studies (ultrasonography, CT, and MRI; Figs. 1–3) and subsequently confirmed at the time of operation and by a histopathological report indicating the presence of neural tissue. The presence of progressive hydrocephalus was determined by an abnormal increase in head circumference, full fontanelle, and increase in ventricular size on imaging studies. Cranial anomalies refer to both CNS and bone anatomy abnormalities. Follow-up information was gathered from outpatient visits to the neurosurgeon or neurologist. Assessments from these outpatient follow-up visits were made regarding motor, visual, hearing, cognitive, and verbal developmental modalities. Children who reached all milestones appropriately were considered to have normal neurological status. Children who did not meet all milestones but could not be differentiated as having mild or moderate delay without data from a validated scale were compiled into 1 group. The designation of severe developmental delay was assigned to children who were nonambulatory and nonverbal. This study was conducted with approval from the Institutional Review Board of Children's Hospital Los Angeles.

Statistical Analysis

Data management and statistical analyses were performed using the program SAS (version 9.3, SAS Institute, Inc.). The independent variables of interest included size of encephalocele in maximal diameter (cm), measured mainly on imaging with a few completed clinically, contents within the encephalocele (neural tissue and/or fluid-filled), microcephaly, presence of associated cranial anomalies, location anterior or posterior to coronal suture, and a seizure disorder. Due to the relatively small sample size, exact logistic regression was used to estimate and test the associations of these independent variables with the two outcomes (presence of hydrocephalus and developmental delay). Each of the independent variables was first modeled separately to estimate its univariate association with the outcome variable. Multivariable exact logistic regression was then used to fit a joint model of independent variables. A variable was considered to be a confounder if it changed the estimated odds ratio (OR) corresponding to any independent variable of interest by more than 15%; statistically nonsignificant factors were excluded from the multivariate model if exclusion did not change the remaining OR estimates by more than 15%. Development or presentation of hydrocephalus and neurological outcome were summarized as ORs, corresponding 95% confidence intervals (CIs), and exact p values.

Results

Patient Characteristics

A total of 70 patients, including 38 girls and 32 boys, were included in the analyses (Table 1). Median age at resection was 2 months (range 0–71 months). Follow-up ranged from 0 to 19 years, with a mean and median follow-up of 3.7 years and 2.2 years, respectively.

Fifty-six patients (80%) presented with a posterior encephalocele, while 14 (20%) presented with an anterior encephalocele. The size of the encephalocele ranged from 0.5 to 23 cm (Fig. 4 upper) in maximal diameter, with a median of 3.1 cm and a mean of 4.0 cm. In Fig. 4 lower we see a patient with massive CNS disorganization in whom there would be a poor chance of survival, and the possibility of cognitive development was nonexistent. With parental concurrence, the patient was not treated and subsequently died. This patient was not included in our series. Forty-seven encephaloceles (67%) contained neural tissue, while in 23 (33%), neural tissue could not be discerned. Eight infants (11%) presented with CSF leaking from the encephalocele, with 1 being infected. Nine patients (13%) developed a seizure disorder following repair. Forty-three patients (61%) had associated cranial anomalies (22 with solely cranial anomalies and 21 with a combination of both cranial and extracranial anomalies), 7 patients (10%) had only extracranial anomalies, while 20 (29%) had no demonstrable abnormalities. The most commonly associated cranial abnormalities included hydrocephalus, Chiari malformation with or without hydrosyringomyelia, agenesis/dysgenesis of corpus callosum, and microcephaly (Table 2). Commonly associated extracranial anomalies included facial dysmorphisms and cleft lip/palate.

Outcome Data

All patients underwent surgical excision and repair of the encephalocele. Postoperatively, 2 patients developed a CSF leak, 1 of whom became infected. Of the 8 neonates with CSF leaking from the encephalocele at birth, 1 was infected and another developed progressive hydrocephalus and required CSF diversion. Seventeen patients (24%) had associated hydrocephalus, with 6 (35%) diagnosed preoperatively, while 11 (65%) developed progressive hydrocephalus postoperatively (Table 1). Fifteen of the patients with hydrocephalus received shunts. One patient was allowed to die without shunt placement and another did not develop progressive hydrocephalus and thus did not require CSF diversion. Three shunt revisions were performed in total. After a median follow-up of 2.2 years, the overall survival rate was 94% (66/70). The 4 patients who died all presented with occipital encephaloceles larger than 5 cm and died of causes unrelated to repair of the encephalocele or a shunting procedure.

In terms of outcome, 14 patients (20%) had severe developmental delay, 28 (40%) had mild/moderate delay, and 28 (40%) were neurologically normal (Table 1). None of the patients with hydrocephalus had a normal neurological outcome; all 17 had some degree of developmental delay, including 11 (65%) with mild/moderate delay and 6 (35%) with severe delay. Two (14%) of 14 patients with an anterior encephalocele developed progressive hydrocephalus in comparison with 15 (27%) of 56 patients with a posterior encephalocele;

the difference in rates was not significant (p = 0.49, Fisher's exact test). Furthermore, 7 (50%) of 14 patients with anterior lesions compared with 35 (63%) of 56 patients with posterior lesions were found to have developmental delay; again, the difference in rates was not significant (p = 0.39, chi-square test).

Correlates of Hydrocephalus and Developmental Delay

Table 3 presents the ORs and 95% CIs from the univariate and multivariate exact logistic regression models for association with hydrocephalus, as well as the prevalence of the independent variables by hydrocephalus outcome. On univariate analysis, the presence of neural tissue, cranial anomalies, encephalocele size of at least 2 cm, seizure disorder, and microcephaly were each positively associated with hydrocephalus. Age at surgery was a confounder as the addition of this variable to the multivariate model altered the estimated ORs of independent factors by more than 15%. Only the encephalocele content variable remained associated with hydrocephalus at a borderline statistically significant level (p = 0.09) in the multivariate model; other variables were retained in the multivariate model as confounders. Because an encephalocele is a rare condition and only 25% of the infants in this sample had hydrocephalus, cranial anomalies, size of encephalocele, and seizure disorder were included in the final model in the interest of estimating effect size rather than testing for statistical significance. These independent factors were positively associated with hydrocephalus in this final model. Adjusting for age at surgery, infants with neural tissue within the encephalocele were 5.8 times more likely (95% CI 0.8–74.0) to present with or develop hydrocephalus after surgery than those with only a fluid-filled encephalocele. Although not statistically significant, children with maximal diameter of the encephalocele of at least 2 cm were 1.7 times as likely (95% CI 0.2–infinity) to present with or develop hydrocephalus. Children with cranial anomalies were 2.4 times as likely (95% CI 0.3–32.7) and children with a seizure disorder were 8.4 times as likely (95% CI 0.7–536.8) to present with or develop hydrocephalus. When microcephaly was added to this multivariable model, it was no longer significantly associated with hydrocephalus (adjusted OR 2.2, 95% CI 0.2–234.7).

Table 4 provides the estimated ORs for associations with developmental delay from univariate and multivariate exact logistic regression models. In univariate models, the presence of cranial anomalies, larger size of encephalocele, hydrocephalus, and microcephaly were positively associated with neurological deficits. In the multivariate logistic regression model adjusted for age at surgery, the presence of hydrocephalus was the most prominent, and only statistically significant, factor (OR 17.2, 95% CI 1.7–infinity). Although not statistically significant, patients with cranial anomalies were 5.4 times as likely (95% CI 0.9–56.7) to exhibit developmental delay. In the multivariate model, encephalocele size was not associated with neurological outcome, and microcephaly was positively but not significantly associated with neurological outcome (OR 6.1, 95% CI 0.5–infinity).

Discussion

Encephaloceles probably occur after neural tube closure, as is evidenced by the cerebral cortex or cerebellar tissue contained within the encephalocele in a majority of cases.[12,15,18]

This CNS herniation may occur 8–12 weeks into gestation at the site of local mesenchymal disruption.[25] While prenatal ultrasonography can be used to detect an encephalocele and the presence or absence of solid tissue within the sac, hydrocephalus is often not present. Various patient series from North America show hydrocephalus is associated with large posterior encephaloceles, although it is more likely to develop following repair of the lesion than be present at birth. In this review we investigate the significant risk factors for both progressive hydrocephalus and developmental delay, while also undergoing a further analysis as to the influence of hydrocephalus on neurological outcome.

In our total cohort of patients, 60% had some degree of cognitive deficit with 40% having a normal neurological outcome. This result is higher than the 16%–31% who were "physically and/or mentally disabled" reported by Tsuchida et al.,[33] Docherty et al.,[11] Date et al.,[9] Macfarlane et al.,[21] and Martinez-Lage et al.[24] Among the hydrocephalic patients in our study, none had a normal neurological outcome. Our review has confirmed previous findings that hydrocephalus is significantly associated with cognitive deficit in patients with encephaloceles.[8,9,19] While patients with posterior encephaloceles had a higher rate of progressive hydrocephalus (15/56) and developmental delay (35/56) than those with anterior lesions (2/14 and 7/14, respectively), there was no statistically significant difference between these rates, confirming the finding by Lo et al.[19] A sample of large reviews is shown in Table 5 for comparison with our results.

The limitations of this study include the retrospective nature of this review, small sample size, and inability to evaluate patients on a uniform cognitive outcome scale. Some infants with normal developmental milestones who were classified as neurologically normal, on longer follow-up may be identified as having learning disabilities, and thus would fall into the mild/moderate developmental delay category.

Conclusions

In multivariate models, the presence of neural tissue was significantly associated with hydrocephalus, and the presence of hydrocephalus was significantly associated with neurological deficit. The location of an encephalocele did not have a statistically significant association with incidence of hydrocephalus or neurological deficit. In contrast to modestly good/fair neurological outcome in children with an encephalocele without hydrocephalus, the presence of hydrocephalus resulted in a far worse neurological outcome. We believe that the information gleaned from this review of our series can help to guide prenatal or neonatal counseling.

Acknowledgments

ABBREVIATIONS

CI   confidence interval

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OR   odds ratio

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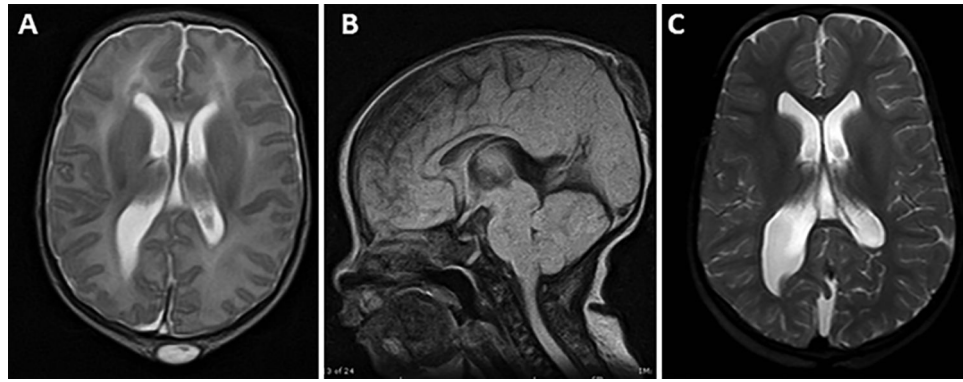
FIG. 1.
Magnetic resonance images obtained in a patient with a small encephalocele containing no neural tissue, no hydrocephalus, no intracranial abnormalities, and a normal development. **A and B:** Axial T2-weighted (A) and sagittal T1-weighted (B) images of a 2-day-old neonate showing a visible mass in the parietal occipital region measuring 1.5 × 2.6 cm. There was an area of dysplastic skin covering the defect without CSF leakage. **C:** At two and a half years of age, an axial T2-weighted image shows no progressive hydrocephalus and the toddler has displayed no apparent neurological defects.
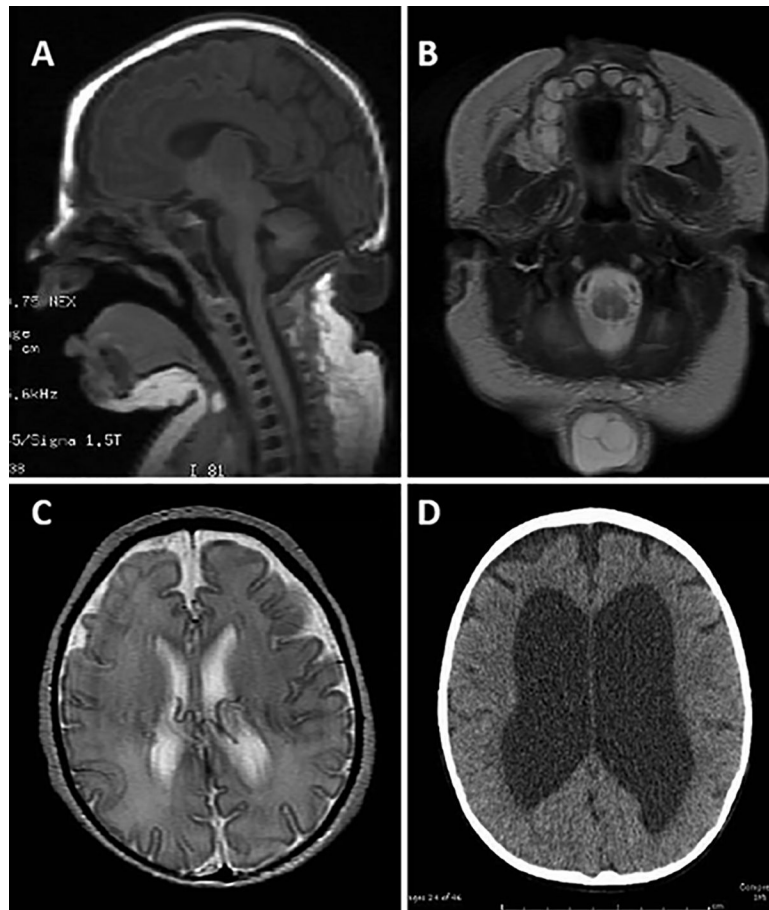
FIG. 2.
Images from a patient with a small encephalocele containing neural tissue, hydrocephalus, no intracranial abnormalities, and a mild/moderate developmental delay. **A and B**: Sagittal T1-weighted (A) and axial T2-weighted (B) MR images of a 6-day-old neonate demonstrate an occipital encephalocele measuring 2.4 cm in maximal diameter containing neural tissue. Her encephalocele was repaired at 11 days of age. **C and D**: Despite normal ventricular size at birth on an axial T2-weighted MR image (C), she developed hydrocephalus as shown on a CT scan 2 months later (D), whereupon a ventriculoperitoneal shunt was placed. A year and a half later, at last follow-up, her cognitive examination found mildly delayed developmental milestones.

FIG. 3.

Images from a patient with a large encephalocele containing neural tissue, hydrocephalus, intracranial abnormalities, and a severe developmental delay. **A and B**: Preoperative T2-weighted sagittal (A) and axial (B) MR images in a neonate born with a large encephalocele (5 cm) containing extensive dysplastic neural tissue with associated microcephaly. She was operated on during the 1st week of life for repair of the encephalocele. Eight days later the child developed a tense fontanelle. **C:** Moderately enlarged ventricles were demonstrated on head ultrasonography and a ventriculoperitoneal shunt was inserted. At 6 months of age, the child displayed severe developmental delay.

FIG. 4.
Photographs of infants with severe encephaloceles. **Upper:** This patient was not initially treated due to the severity of her malformation, but as she survived her encephalocele was repaired at 6 months of age and a ventriculoperitoneal shunt was placed to control progressive hydrocephalus. **Lower:** Due to the severity of this malformation and the poor prognosis, this neonate was not treated and allowed to die (not included in our series).

TABLE 1.

Patient characteristics

Variable	Value
No. of patients	70
Sex (%)	
Male	32 (46)
Female	38 (54)
Age at surgery (mos)	
Range	0–71
Median	2
Mean	8.5
Hydrocephalus (%)	17 (24)
Presented	6
Developed	11
Neurological outcome (%)	
Normal	28 (40)
Mild/moderate delay	28 (40)
Severe delay	14 (20)
Follow-up (yrs)	
Range	0–19
Mean	3.7
Median	2.2

TABLE 2.

Associated cranial and extracranial anomalies

Anomaly	Value
Cranial	
Microcephaly	13
Agenesis/dysgenesis of corpus callosum	10
Chiari malformation	6
Schizencephaly	3
Arachnoid cyst	3
Walker-Warburg malformation	2
Dandy-Walker malformation	2
Holoprosencephaly	2
White matter hypoplasia	1
Sagittal synostosis	1
Extracranial	
Facial dysmorphisms	9
Cleft lip/palate	6
Hydrosyringomyelia	3
Aplasia cutis congenita	2
Aplasia of bilateral thumbs	1

TABLE 3.

Associations with hydrocephalus outcome according to exact logistic regression

	Hydrocephalus		Univariate		Multivariate[*]	
Variable	**Yes (n = 17)**	**No (n = 53)**	**OR (95% CI)**	**p Value**	**OR (95% CI)**	**p Value**
Neural tissue present	15 (88%)	32 (60%)	4.8 (1.0–47.8)	0.06	5.8 (0.8–74.0)	0.09
Cranial anomalies present	15 (88%)	28 (53%)	7.0 (1.4–69.6)	0.01	2.4 (0.3–32.7)	0.63
Size 2 cm	17 (100%)	39 (74%)	8.2[†] (1.2–infinity)	0.02	1.7[†] (0.2–infinity)	0.68
Posterior location	15 (88%)	41 (77%)	2.2 (0.4–22.3)	0.55		
Seizure disorder	7 (41%)	2 (4%)	16.8 (2.7–188.5)	0.001	8.4 (0.7–536.8)	0.14
Microcephaly	7 (41%)	6 (11%)	5.63 (1.29–26.10)	0.02		

[*]
Adjusted for age at surgery.

[†]
Indicates a median unbiased estimate.

TABLE 4.

Associations with developmental delay according to exact logistic regression

Variable	Developmental Delay		Univariate		Multivariate*	
	Yes (n = 42)	No (n = 28)	OR (95% CI)	p Value	OR (95% CI)	p Value
Cranial anomalies present	32 (76%)	11 (39%)	4.3 (1.4–13.8)	0.008	5.4 (0.9–56.7)	0.07
Size 2 cm	38 (90%)	18 (64%)	5.1 (1.3–25.6)	0.02	1.2 (0.15–11.23)	1.0
Hydrocephalus	17 (40%)	0 (0%)	$25.3^{†}$ (4.0–infinity)	<0.0001	$17.2^{†}$ (1.7–infinity)	0.01
Neural tissue present	28 (67%)	19 (68%)	1.2 (0.4–3.8)	0.87		
Posterior location	35 (83%)	21 (75%)	1.7 (0.4–6.4)	0.58		
CSF leak	8 (19%)	2 (7%)	3.0 (0.5–31.5)	0.30		
Seizure disorder	8 (19%)	1 (4%)	6.2 (0.8–292.1)	0.12		
Microcephaly	13 (31%)	0 (0%)	$17.2^{†}$ (2.7–infinity)	0.0009	$6.1^{†}$ (0.5–infinity)	0.2

*
Adjusted for age at surgery.

†
Indicates a median unbiased estimate.

TABLE 5.

A review of comparable series showing the variability in reports on the effects of various factors on long-term neurological outcome in children born with an encephalocele

Authors & Year	No. of Patients	Factors Affecting Long-Term Neurological Outcome		Comments
		Hydrocephalus	Neural Tissue	
Lo et al., 2008	85	Yes	Yes	
Bui et al., 2007	44	Yes		Series of cranial vault encephaloceles
Martinez-Lage et al., 1996	46	Yes	No	
Date et al., 1993	24	Yes	Yes	Series of occipital encephaloceles
Shokunbi et al., 1988	58	No	No	Series of sincipital encephaloceles
Mealey et al., 1970	65	No	Yes	