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Basal encephalocoele: imaging and exposing the hernia

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SUMMARY. **Basal encephalocoeles are rarely reported anomalies. Eight cases seen by one unit manifested external facial features and internal cerebral anomalies characteristic of the individual encephalocoele subgroups. CT and MR imaging delineates the anatomy of the skeletal defect and the associated cerebral abnormalities. Such imaging of cases of median cleft face syndrome may identify previously unsuspected basal encephalocoeles.**

Transcranial correction with increased exposure, if needed, by the technique of facial bipartition has been performed in five cases.

Basal encephalocoele, a developmental herniation of cerebral and meningeal tissue through a defect in the cranial base, has long been identified as a rare cause of soft tissue protrusion into the orbit, nasal cavity, nasopharynx or paranasal sinuses. Classification of basal encephalocoeles into subtypes based on the anatomical point of exit—trans-ethmoidal (TEE), spheno-ethmoidal (SEE), spheno-maxillary, spheno-orbital, and trans-sphenoidal (TSE)—is well accepted.[1] Similarly, the pattern of associated craniofacial, cerebral and ophthalmic anomalies with particular subtypes is documented and provides some opportunity to speculate on aetiology.[2–6] Theories ranging through failure of ossification centres in the sphenoid,[7] persistence of the craniopharyngeal canal and failure of closure of the anterior neuropore,[8] and anomalous induction of mesenchymal tissues or failure of fusion of the neural folds into a neural tube at this point have all been postulated as causative of basal encephalocoeles.[9]

The concealed, occult location and apparently recondite nature of the basal encephalocoele has also provided a continuing challenge to both diagnosis and management. The advent of complex techniques of three dimensional imaging and surgical exposure of this region has largely overcome these problems, permitting earlier accurate diagnostic delineation and the potential for definitive correction of both the primary encephalocoele and the secondary growth disturbances it may induce.

This study reviews the experience of the Australian Craniofacial Unit in managing basal encephalocoeles between 1986 and 1992.

Patients

A review of the records of the Australian Craniofacial Unit revealed eight patients with the diagnosis of basal encephalocoele. Six underwent a detailed multidisciplinary assessment in the unit before proceeding to surgical correction, while the remaining two were reviewed in overseas clinics but declined further treatment.

Complex radiologic investigation included cephalometric radiography, axial and coronal 2-D CT scanning with 3-D CT reconstruction and, in the last four patients, MRI scanning.

Transcranial correction, performed at the earliest opportunity so as to minimise any distorting influence on craniofacial growth, has been favoured in this unit.

Results

In this series the distribution of encephalocoele type is recorded in Table 1. The TEE tended to present later in life, when compared to the TSE. The associated facial anomalies in the TEE group were minimal, with one case demonstrating mild hypertelorism and another a cleft lip. The patient with SEE (Figs 1, 2) and three of the TSE group all had marked hypertelorism, cleft lip and palate, these features of the median cleft face syndrome precipitating their early diagnosis (see Table 2).

Agenesis of the corpus callosum (see Table 3) was only seen in the sphenoidal encephalocoeles, conforming to the view that only TSE are seen in association with the median cleft face syndrome.[2,6] CT scan and MRI scan revealed a small TSE in one case with multiple rare craniofacial clefts, bilateral nasal probosci and unilateral microphthalmus, thought to be an example of the amniotic band sequence. At the time of transcranial correction of the craniofacial clefts, no encephalocoele was identified, although

Table 1 Distribution of basal encephalocoele type ($n = 8$)

Trans-ethmoidal (TEE)	3
Spheno-ethmoidal (SEE)	1
Spheno-maxillary	0
Spheno-orbital	0
Trans-sphenoidal	4

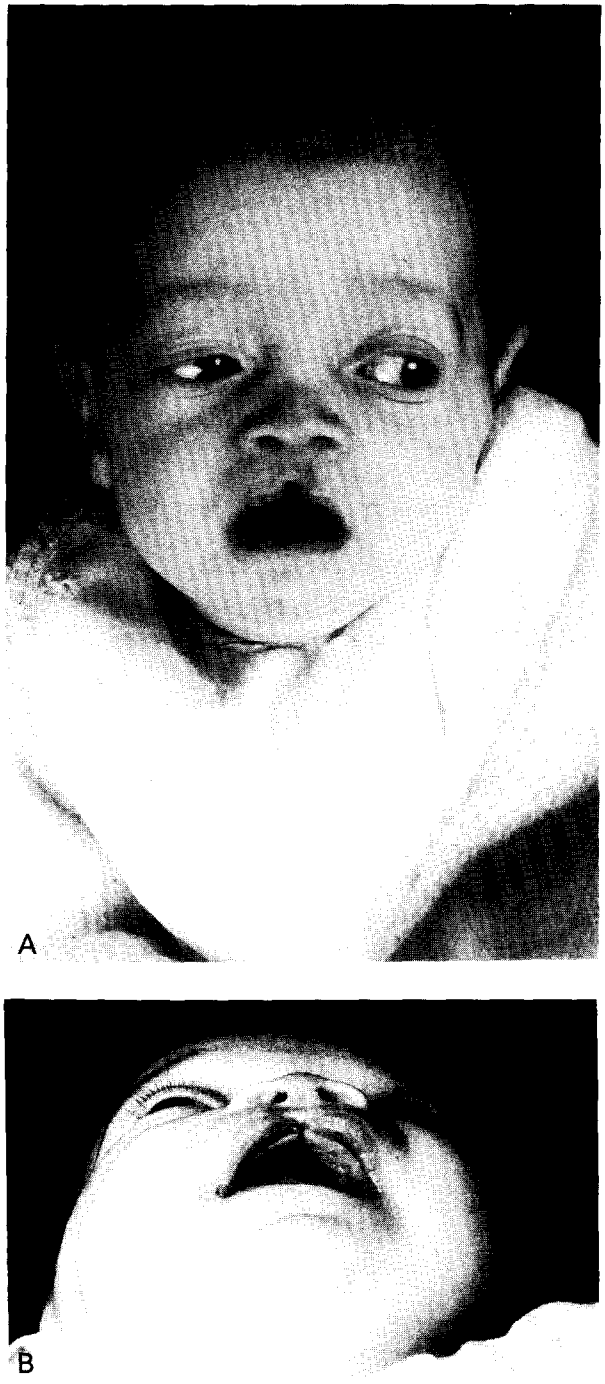

Fig. 1

Figure 1—(A) Four month old child with a spheno-ethmoidal meningo-encephalocoele demonstrating median cleft lip and (B) median cleft alveolus.

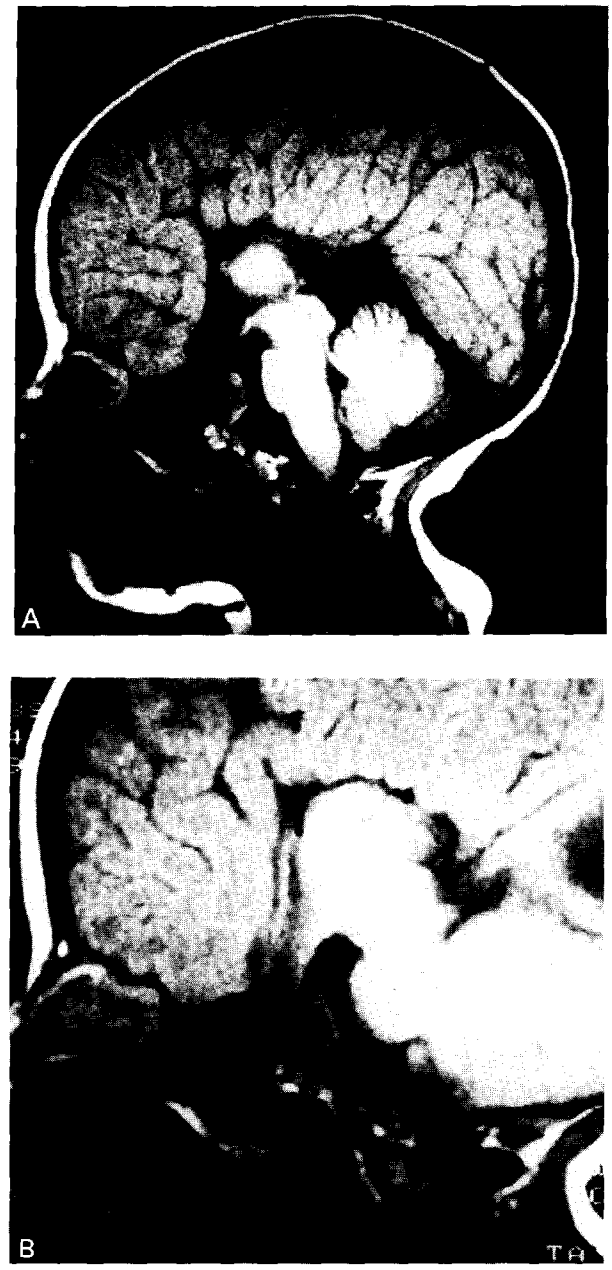

Fig. 2

Figure 2—(A, B) MRI scan of four month old child with a spheno-ethmoidal meningo-encephalocoele, sagittal views.

dissection may not have been carried sufficiently posteriorly to identify brain herniation in the region of the sphenoid sinus and pituitary fossa (Table 4).

Presentation in the TEE group in two cases was to an otorhinolaryngologist for persistent rhinorrhoea. One had experienced several episodes of meningitis since a neonate, with eventual insertion of a ventriculo-peritoneal shunt for treatment of hydrocephalus. The remaining case of TEE presented with symptoms of panhypopituitarism (see Table 2).

Prior to surgery, hypothalamic-pituitary function was assessed by hormone assays. This was repeated postoperatively and found to be unchanged. One patient with SEE was found to have pan-hypopituitarism 18 months post surgery and is responding well to replacement therapy.

Surgical correction of the encephalocoele was performed in five cases (Table 4). The TEE were approached via a bifrontal craniotomy with reduction of the herniated brain and meningeal coverings and bone grafting of the cranial base defect.

The more posteriorly placed SEE and TSE were approached using a combination of the above trans-cranial approach and facial bipartition. This permitted concomitant correction of both the encephalocoele and the hypertelorism in patients as young as 4 months of age, with an uncomplicated postoperative course. In

Table 2 Patient characteristics

Case	*Encephalocoele type*	*Age at presentation and reason*	*Facial morphology*	*Panhypopituitarism*
1.	TEE	6 years (Panhypopituitarism)	Cleft lip	Yes
2.	TSE	Birth (Median cleft face)	Cleft lip, palate Hypertelorism	Yes
3.	TEE	10 years (Rhinorrhoea)	Hypertelorism	No
4.	TSE	Birth (Median cleft face)	Cleft lip, palate Hypertelorism	No
5.	SEE	Birth (Median cleft face)	Cleft lip, palate Hypertelorism	No
6.	TEE	3 years (Rhinorrhoea)	Normal	No
7.	TSE	Birth (Median cleft face)	Cleft lip, palate Hypertelorism	Yes
8.	TSE	1 year (Incidental finding on MRI)	Tessier clefts Hypertelorism	No

Table 3 Neurological features

Case	*Encephalocoele type*	*Corpus Callosum*	*Meningitis*	*Mental retardation*
1.	TEE	Present	No	No
2.	TSE	Absent	Post-op	Yes
3.	TEE	Present	Prior to diagnosis	No
4.	TSE	Present	No	No
5.	SEE	Absent	No	Developmental delay
6.	TEE	Present	Prior to diagnosis	Yes
7.	TSE	Unknown	No	No
8.	TSE	Present	No	No

Table 4 Surgical intervention in basal encephalocoele

Case	*Encephalocoele type*	*Surgery*	*Age at surgery*
1.	TEE	No surgery	
2.	TSE	Transcranial correction (not at ACPU)	6 years
3.	TEE	Transcranial correction	10 years
4.	TSE	Facial bipartition Transcranial correction	2 years
5.	SEE	Facial bipartition Transcranial correction	4 months
6.	TEE	Transcranial correction	3 years
7.	TSE	Transcranial correction	1 year
8.	TSE	Transcranial correction of Tessier clefts Exploration for basal encephalocoele	1 year

these cases bone graft and a galeofrontalis flap was interposed in the region of the defective cranial base (Figs 3, 4).

Discussion

Basal encephalocoeles occur at a reported rate of 1 in 35,000 live births. Their occult location, and past difficulties in both imaging and exposing this area, have undoubtedly contributed to underdiagnosis. Whilst localisation to involvement of either the ethmoid or sphenoid bones may help in determining the symptomatology of how the encephalocoele will present, it does not necessarily prevent confusion with more common conditions, and thus misdiagnosis.

Heinecke in 1882 was reported by Fenger[10] to have originally classified basal encephalocoeles as sphenopharyngeal, spheno-orbital or spheno-maxillary in type. Subsequently Gisselson[1] expanded the classification, by labelling them according to the position of exit of the encephalocoele.

The TEE presents the more challenging diagnostic dilemma as associated facial and cerebral anomalies, which may act as a marker for the encephalocoele, are much less frequent. The presence of an intranasal

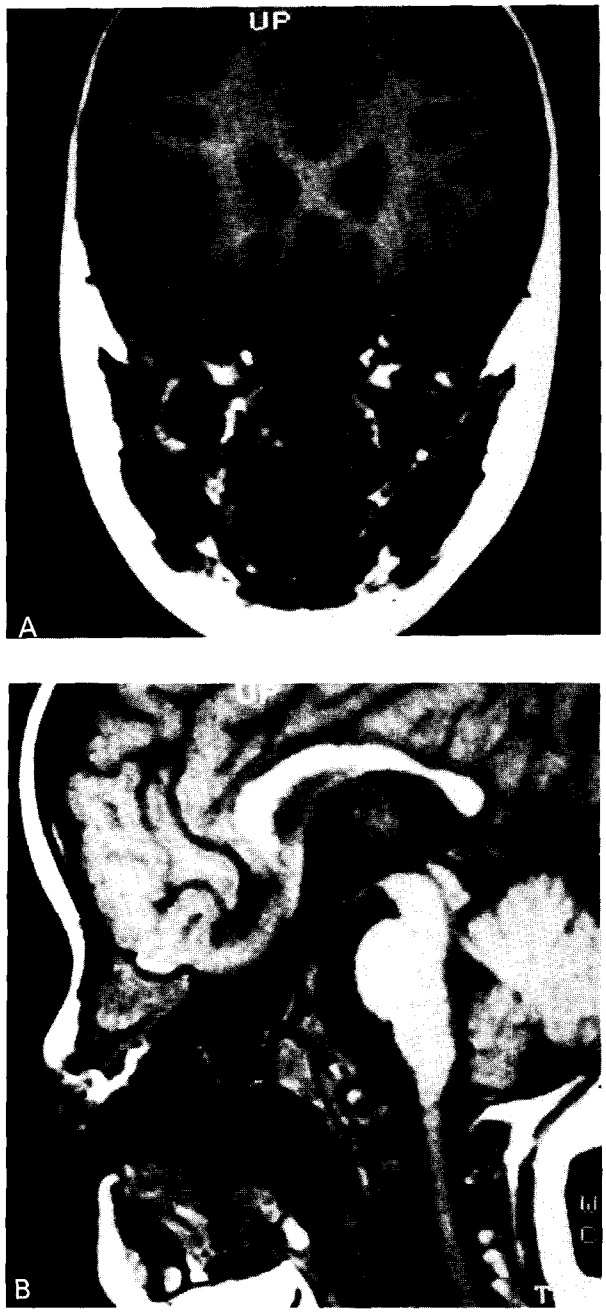

Fig. 3

Figure 3—(A) MRI scan of 2 year old child with a trans-sphenoidal meningo-encephalocoele, coronal view. (B) MRI scan of 2 year old child with a trans-sphenoidal meningo-encephalocoele, sagittal view.

tumour, which pulsates synchronously with the pulse or respiration, with symptoms of nasal obstruction from birth, requires appropriate assessment and imaging with MRI to exclude a TEE. Failure to consider this diagnosis risks the initiation of inappropriate treatment complicated by cerebrospinal fluid leak and meningitis.[11–13]

In contrast, the encephalocoeles which are transmitted through the sphenoid bone will most commonly have co-existing anomalies of the face, optic system and brain, corresponding to the median cleft face syndrome.[14] The facial manifestations, hypertelorism, broad nasal root, median cleft nose, median cleft lip and maxilla and cranium bifidum occultum frontale are much more arresting and when seen require exclusion of an associated basal encephalocoele. This is particularly so where transcranial correction of the hypertelorism is envisaged. In this group there is also an increased incidence of anomalies of the optic disc, optic nerves, lipomas or agenesis of the corpus callosum and concomitant widening of the lateral ventricles. These associated findings all add to the speculation and uncertainty regarding aetiology.

Importantly, these more posteriorly located basal encephalocoeles may be associated with hypothalamic-pituitary dysfunction[15] and indeed knowing whether these regions are part of the herniated cerebral tissue is essential prior to any surgical intervention.

Detailed 3-D CT reconstruction of the cranial base allows one to view the skeletal outlines of the hernia. Bony marginal hypoplasia is not evident—all bony elements are present, although displaced, where the encephalocoele is of significant size. These techniques, when used routinely in the major midline craniofacial anomalies, such as median cleft face syndrome, will reveal the hitherto unsuspected basal encephalocoele. Visualising the anatomy of these disturbances in the cranial base then provides the opportunity to plan surgical correction appropriately, whether this be by hernia reduction and bone graft alone, or in combination with extensive craniofacial osteotomies.

Quantitative analysis, particularly in those late presenting cases, may in the future allow assessment of the growth disturbing influence of the encephalocoele on midfacial growth.

Conventional exposure by frontal craniotomy has been successful for ethmoidal encephalocoeles, presenting as they do in the older age group. In the light of the potential for earlier diagnosis, with the advent of improved imaging, there are no contraindications to definitive early correction. Reduction of the anomalous cerebral tissue via this route then permits closure and sealing of the cranial base defect with bone graft and a vascularised galeofrontalis flap. Any inadvertent penetration of the nasal cavity is then separated by vascularised tissue from direct communication with the intracranial space.

Encephalocoeles originating in the region of the sphenoid bone are more taxing both in terms of exposure and requirements for preservation of vital functional cerebral tissue—the pituitary–hypothalamic axis. Additionally, their association with the median cleft face syndrome introduces the extra dimension of hypertelorism correction. Facial bipartition, performed by convention at approximately five years of age, facilitates both encephalocoele exposure and correction of hypertelorism, with minimal risk of damage to the midfacial unerupted secondary dentition. Where the extra mobilisation necessary for hypertelorism correction is not required, facial bipartition may be employed even earlier—in this series as young as four months of age. Mini- or microplate fixation provides precise stable reconstruction of the face, and calvarial bone the ideal donor site for graft material to occlude the cranial base defect.

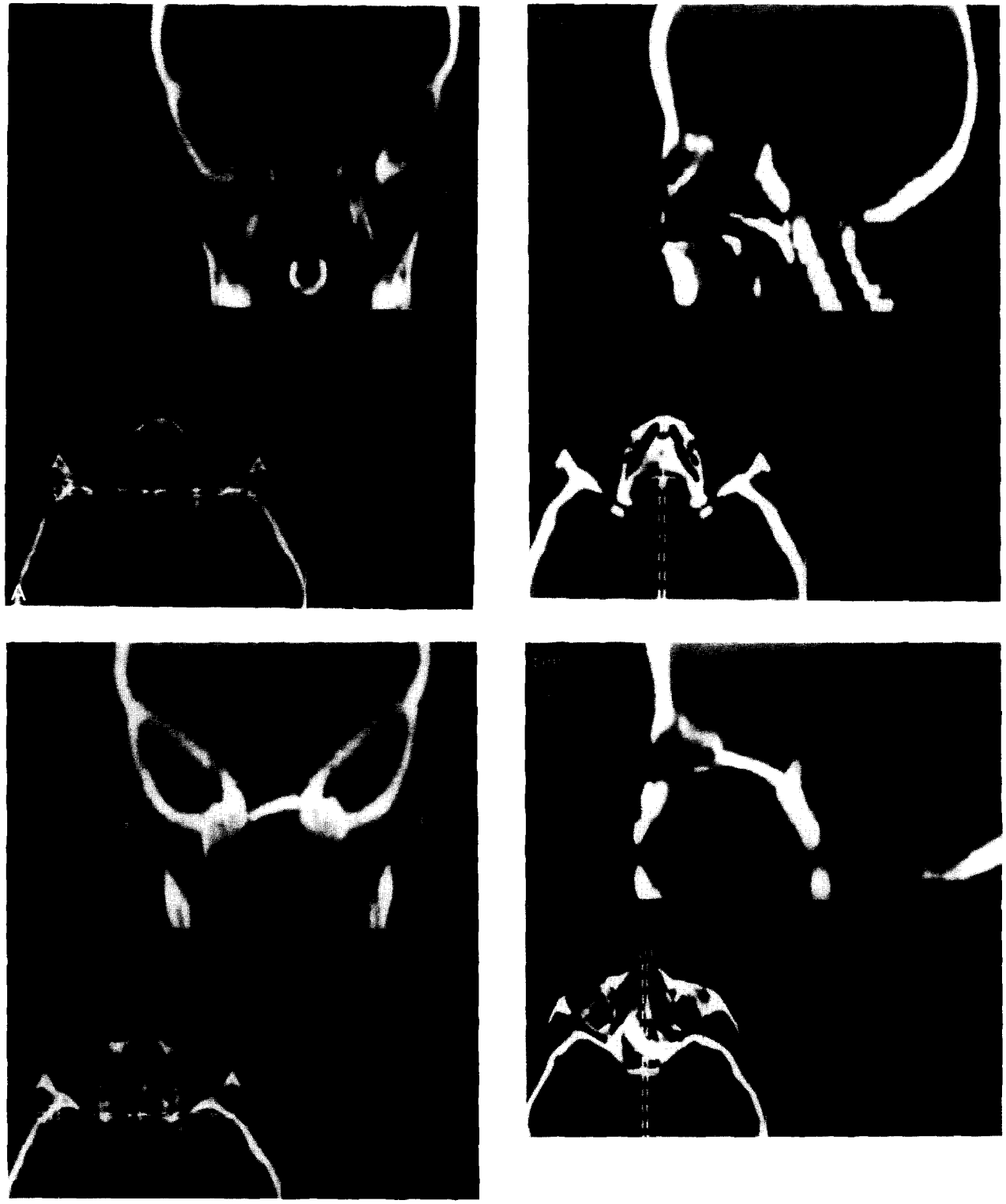

Fig. 4

Figure 4 (A) CT scan of 2 year old child with a trans-sphenoidal meningo-encephalocoele, coronal view. (B) CT scan of 2 year old child with a trans-sphenoidal meningo-encephalocoele, sagittal view. (C) CT scan of 2 year old child after correction of trans-sphenoidal meningo-encephalocoele, demonstrating the bone graft *in situ*, coronal view. (D) CT scan of 2 year old child after correction of trans-sphenoidal meningo-encephalocoele, demonstrating the bone graft *in situ*, sagittal view.

Future observations of facial growth following cranial base reconstruction in these cases are planned.

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