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Original Article

A Clinicopathological Classification Of Encephalocoeles Based On 207 Patients ☆,☆☆

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ABSTRACT

Introduction: Encephalocoeles are relatively rare congenital abnormalities. There have been a few classifications of encephalocoeles, but these are predominantly anatomical. A more clinical classification system would assist in planning treatment, surgical procedures and assessing outcomes.
Materials and Methods: All encephalocoeles presenting at the Craniofacial Unit at Inkosi Albert Luthuli Central Hospital were reviewed. There were 207 patients with 224 encephalocoeles. The clinical presentation and CT findings were analysed and used to group these encephalocoeles.
Results: There were five distinct groups with some having subgroups.

1. Cranial (n= 43). These were located on the calvarium and were subdivided into subgroups according to their anatomical location. They are occipital, parietal, frontal, temporal and acrania.
2. Nasal (n= 122). These were located in the nasal region and were classified into two large subgroups (supranasal and infranasal) depending on whether the pathway and defect were above or below the nasal bones.
3. Orbital (n= 21). These presented with the displacement of the globe and were subdivided into two subgroups: anterior and posterior.

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4. Basal (n= 11). The pathway of these encephalocoeles was through the floor of the anterior cranial fossa often with no visible deformity of the face.
5. Cleft related (n= 27). The pathway of these encephalocoeles was through an existing craniofacial cleft.

Conclusion: This classification system demonstrated good clinicopathological correlation. This allowed one to better appreciate the pathway and assess concomitant deformities. It also directed one to plan the procedure and detail the surgical corrections required to produce satisfactory outcomes.

An encephalocoele is a relatively rare congenital abnormality with herniation of the brain and meninges outside of the cranial cavity. The incidence varies greatly and seems to have a racial and geographical predilection. It is more common in the Asian and African races and relatively rare in the Caucasian group. The reported incidence is between 0.8 and 3.0 per 10 000 live births.

It may be an isolated finding, but more often than not, there are other associated abnormalities. These anomalies may be in the central nervous system, face, limbs, cardiac, abdominal or genitourinary systems.

Encephalocoeles can have a range of clinical presentations and can form a small lump to grotesque craniofacial deformities. In order to understand encephalocoeles and help in communication, classification systems have been proposed. The most popular classification is that of Suwanela and Suwanela which was reported in 1972.[1] This was based on 12 cases of encephalocoeles which were all post-mortem cases. These were all sincipital encephalocoeles. They classified it into five broad groups with 17 possible subgroups. The five groups were occipital, cranial vault, frontoethmoidal, basal and cranioschisis. At that time, the ability to image encephalocoeles was rudimentary compared to the CT and MRI scans that are available now. Isotope cisternography, ventriculography and air studies were some of the methods used to delineate encephalocoeles.[2,3] These were obviously not very accurate in detailing the pathway and defining the soft tissue and bone abnormalities. There are multiple shortcomings of this classification. Firstly and probably the most significant is that there is little or no clinicopathological correlation to categorize the appearance according to the type of encephalocoele. Secondly, there are some anatomical inaccuracies. One of the frontoethmoidal subgroups is referred to as 'naso-orbital'. In such cases, the nasal bones are not involved, and the nose may be completely normal. Because they had 12 cases and enumerated 17 possible types of encephalocoeles, some of the ones described were regarded as 'theoretical' encephalocoeles. These have now been repeatedly described in the literature. The final limitation of the classification is that it does not take into account newer types of encephalocoeles that have since been described.

The Australian Craniofacial Unit has proposed a classification based on their experience.[4,5] The second paper is based on 23 patients with frontoethmoidal encephalomeningoceles, and they termed it the facial deformity, external bone defect, exit pathway and malformation of the brain (FEEM) classification.

Because of its rarity, most reports on encephalocoeles consist of a small number of patients. A large series of 400 cases has been reported by Arifin (Indonesia), 133 by Mahapatra (India) and 108 by Mahatumarat (Thailand), which were all mainly frontoethmoidal ones.[6–8] Shokunbi describes 57 occipital encephalocoeles in a Nigerian population.[9] Most of these studies focused on the surgical outcomes rather than the analysis of the subtype of the encephalocoele. The large series of cases reported by Arifin found that 87% of the cases were of the nasoethmoidal type with or without a naso-orbital component.

Mahatumarat proposed a modification of the Suwanela classification and had five groups which were nasofrontal, nasoethmoidal, naso-orbital, combined and abortive.

We would like to report on our experience of 207 patients with 224 encephalocoeles seen in our craniofacial unit. We propose a new classification of encephalocoeles that has clinicopathological correlation, takes into account newly described encephalocoeles, has simple and less confusing terminology and will allow better communication amongst craniofacial surgeons.

Materials and Methods

The charts of the Craniofacial Unit at Inkosi Albert Luthuli Central Hospital were reviewed, which included patients first seen at other hospitals (Grey's and Wentworth) where the unit was previously based. All patients seen at the craniofacial clinic with a diagnosis of encephalocoeles had their charts reviewed. All patients with CT scan evidence of herniation of the brain and meninges outside the cranial cavity were included. Exclusion criteria were those patients who did not have CT imaging performed or those with no evidence of an encephalocoele on review of the CT scan. It must be noted that some patients, especially those with occipital encephalocoeles, did not present to the craniofacial clinic but were treated in the neurosurgery clinic. There were 207 patients with 224 encephalocoeles, 98 males and 109 females. The predominant race group was the Black population. The ages ranged from 1 day to 6 months. All patients had a CT scan. A nomenclature system was devised whereby the clinical presentation, features and CT scan findings allowed us to categorize these patients into groups. Some of these patients had leaking encephalocoeles, and these were operated on immediately. Those patients who had tenuous skin cover and imminent cerebrospinal fluid (CSF) leaks were also operated on an urgent basis. The other patients were operated on an elective basis at around 3 months of age. Based on the findings, different surgical approaches and strategies were used to excise the encephalocoele and reconstruct the craniofacial skeleton. The surgery aimed to excise the encephalocoele, achieve watertight dural closure, bone graft the defect, correct craniofacial abnormalities, obviate or minimize any visible scars and correct soft tissue deformities such as canthoplasty.

Results

There were 224 encephalocoeles in 207 patients. These encephalocoeles were categorized into groups and subgroups (Table 1). Based on this, the proposed classification has five broad groups: cranial, nasal, orbital, basal and cleft related.

Cranial

This group of encephalocoeles was located in the cranium and named according to their anatomical site. There were 43 patients in this group. The most common ones were the occipital[15] and frontal.[15]

a. Occipital.[15] These presented as a single lump or one with multiple lobes. They were usually in the midline. The size varied from small to ones that almost mirrored the size of the cranium and sometimes larger.
b. Parietal.[6] These were commonly on one side rather than midline.
c. Frontal.[15] These are presented between the frontal bones or on one side.
d. Temporal.[1] These were usually unilateral presenting as a mass.
e. Acrania.[5] There were a few cases where there was near-total absence of the cranial vault bones. These patients usually had severe brain deformities.

Fig. 1 demonstrates the preoperative views of an encephalocoele in the parieto-occipital region.

Nasal

These were all encephalocoeles which presented in the nasal region. This was the most common group and comprised 122 patients. There were 3 groups that we identified that had distinct clinical features that correlated with the tract and pathway of the encephalocoele.

Table 1
Groups, subgroups and number of encephaloceles.

	Number	Subtotal
Cranial:		
Frontal	15	**43**
Occipital	15	
Parietal	6	
Frontoparietal	1	
Temporal	1	
Acrania	5	
Nasal:		
Supranasal	68	**122**
Infranasal	53	
Intranasal	1	
Obital:		
Anterior	19	**21**
Posterior	2	
Basal:		
Ethmoidal	8	**11**
Trans-sphenoidal	2	
Sphenomaxillary	1	
Facial cleft:		
1, 13	15	**27**
0, 14	4	
10	5	
2, 12	2	
9	1	
Grand total		**224**

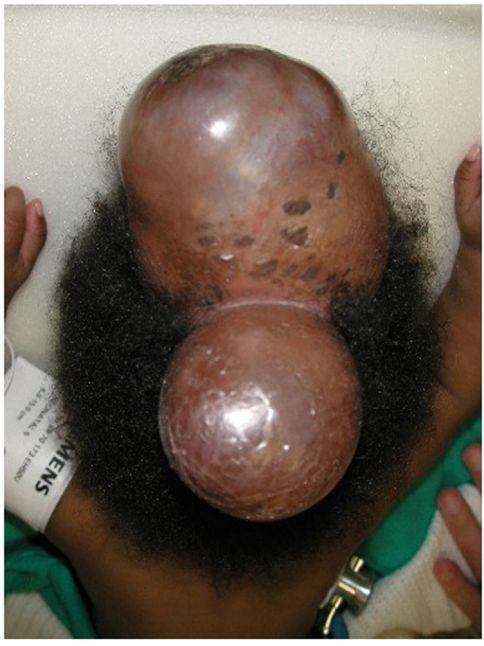

Fig. 1. a. Preoperative view of a parieto-occipital encephalocoele.

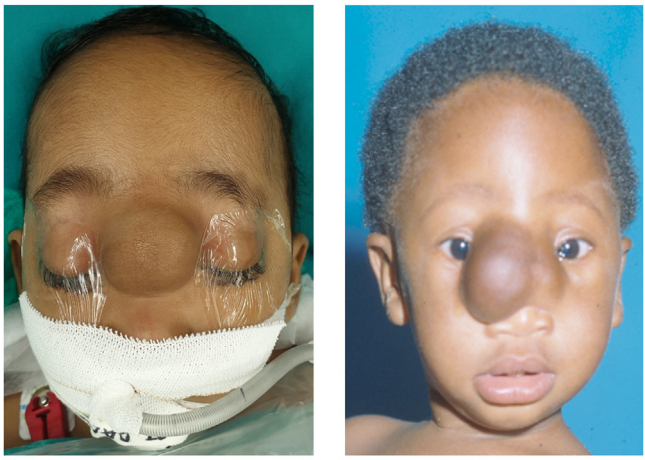

Fig. 2. a. Supranasal encephalocoele. b. Infranasal encephalocoele.

Supranasal

After exiting from the base of the anterior cranial fossa, this encephalocoele exited from the face *above* the nasal bones. The presentation was usually that of a symmetrical midline mass whose maximal diameter was above the medial canthus. There was no associated telecanthus. The nasal bones were displaced in an inferior and posterior direction. There were 68 patients in this group.

Infranasal

This encephalocoele exited from the face *below* the nasal bones. This presents as a bilateral asymmetrical lump whose maximal diameter was below the medial canthus. Telecanthus was common, and there was medial canthus displacement. The nasal bones were displaced in a superior and anterior direction. There were 53 patients in this group.

Fig. 2a, b shows the typical clinical presentation of a supranasal and infranasal encephalocoele. Fig. 3a, b demonstrates the difference in nasal bone displacement with the supranasal and infranasal groups.

Intranasal

This is the least common nasal group (n=1), and the encephalocoele presents as a mass behind the nasal bones, causing it to balloon out.

Orbital

After exiting the anterior cranial fossa, these encephalocoeles exit into the orbit. There are two distinct groups that are seen.

Anterior orbital

These encephalocoeles usually exit between the lacrimal and nasal bones. They present as a mass in the medial canthal region and usually displace the globe laterally and superiorly. There may be displacement of the medial canthus. There were 15 patients in this group. Four of these patients had bilateral encephalocoeles, giving a total of 19 encephalocoeles. Fig. 4 demonstrates the presence of an anterior orbital encephalocoele and the CT images.

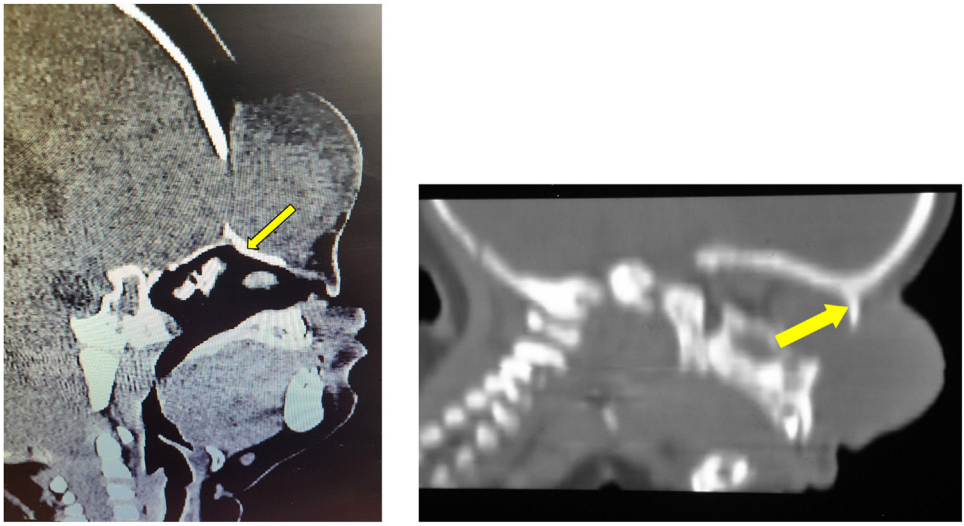

Fig. 3. a. CT scan of supranasal encephalocoele showing nasal bone displacement. b. CT scan of infranasal encephalocoele showing nasal bone displacement.

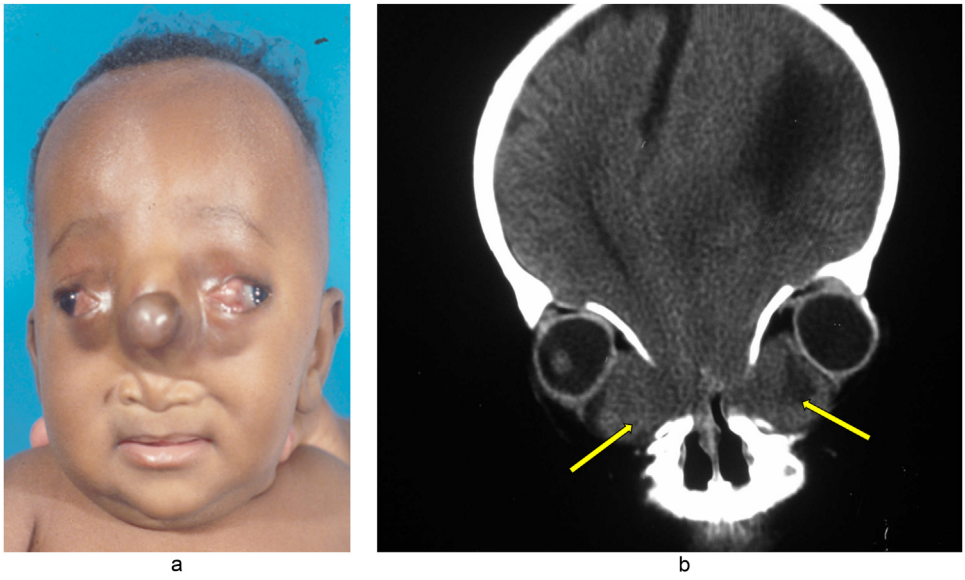

Fig. 4. a. Patient with supranasal and bilateral anterior orbital encephalocoeles. b. CT scan with arrows depicting encephalocoeles.

Posterior orbital

These encephalocoeles come through one of the orbital foramina, the roof of the orbit or the medial orbital wall. They tend to displace the globe anteriorly and present with a unilateral exophthalmos. The foramina that can be the pathway are the anterior ethmoidal, posterior ethmoidal, superior orbital fissure, inferior orbital fissure and optic foramen. There were two patients in this group.

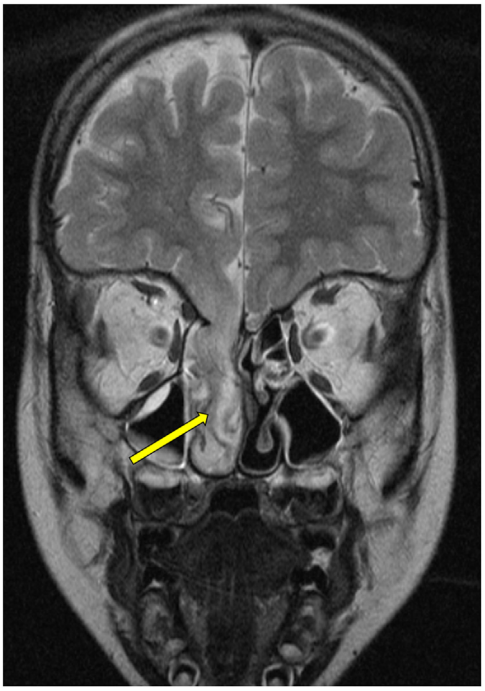

Fig. 5. MRI scan demonstrating an ethmoidal encephalocoele.

Basal

These encephalocoeles herniate through the floor of the anterior, middle or posterior cranial fossae. In the anterior cranial fossa, these are ones that do not present in the nasal or orbital regions. The herniation occurs through the ethmoidal, sphenoidal or temporal bones.[10–17] After herniation, these encephalocoeles can either be occult or present as a visible mass in the lateral facial region.[16–19] The occult ones may present as a mass once they are big enough. These may appear in the auditory canal, nasal cavity, sinuses or posterior pharynx. The occult ones may leak CSF and present with meningitis.[20–23] One needs to have a high index of suspicion and order a CT or MRI scan to make the diagnosis. Sometimes a basal encephalocoele may present as a mass in cleft palate patients. One must resist the temptation to perform a biopsy prior to scanning the patient. There were 11 patients in this group. The majority of these were ethmoidal[8] with two trans-sphenoidal and one sphenomaxillary. Fig. 5 demonstrates the MRI scan of an ethmoidal encephalocoele.

Facial cleft

One may get encephalocoeles associated with the northbound Tessier clefts which are the 8–14 clefts.[24,25] The most common ones are the ones that are paramedian and represent the 11–14 facial clefts which are often associated with hypertelorism. These encephalocoeles may be unilateral or bilateral. The encephalocoele associated with a facial cleft 10 presents as a mass in the middle of the eyebrow. There were 25 patients in this group with 27 encephalocoeles. The most common ones were those associated with a 1, 13 facial cleft (n=15). Four were associated with a 0, 14 cleft and 5 with a 10 cleft. Fig. 6 demonstrates a facial cleft 10 with an accompanying encephalocoele.

One may get a combination of encephalocoeles. These are most commonly seen in the nasal and orbital groups. Five patients had the combination of infranasal and anterior orbital encephalocoeles, whilst four had supranasal and anterior orbital ones.

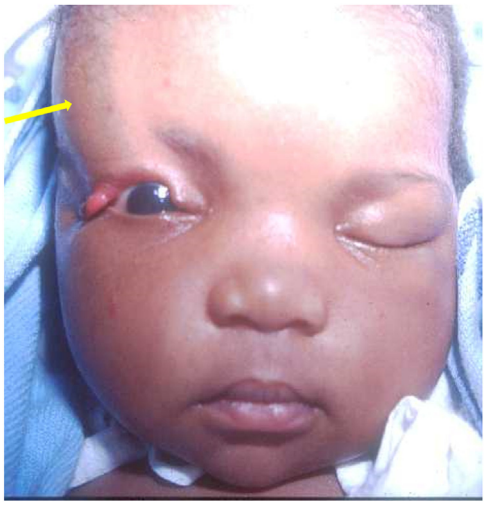

Fig. 6. Patient with a facial cleft 10 showing defect in the right upper eyelid and an encephalocoele.

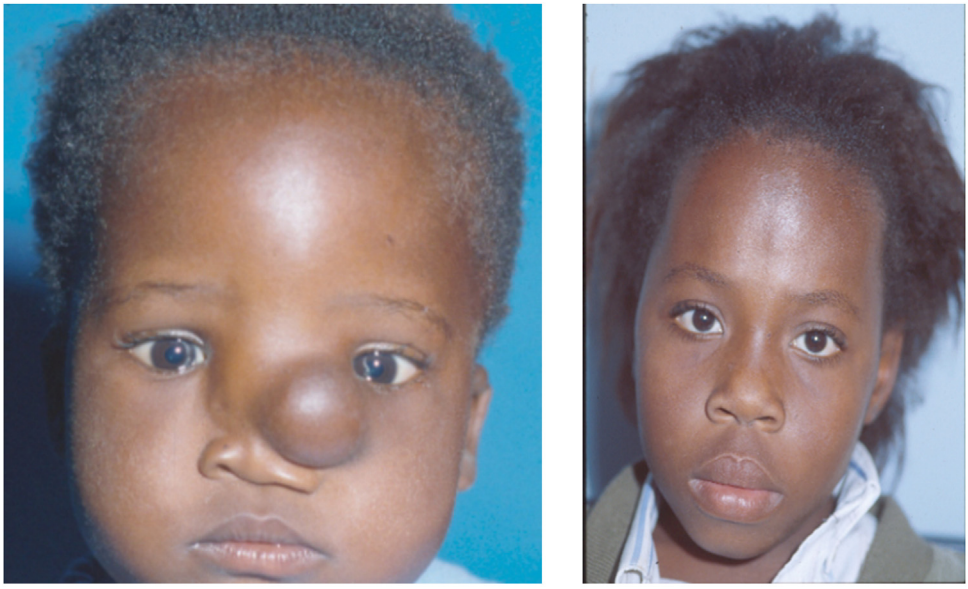

Fig. 7. a. Patient aged 15 months with an infranasal encephalocoele. Coronal and left paranasal incision, frontal craniotomy, bone graft to defect, medial canthoplasty. b. 3-year postoperative view.

These patients were usually operated on at the age of 3 months. If there was a leaking encephalocoele or an imminent leak, this was treated as an emergency, and surgery was performed as soon as possible. The rationale was that a CSF leak runs the risk of an infection, and the onset of ventriculitis usually has a fatal outcome.

Figs. 7–9 show pre- and postoperative views of patients.

Fig. 8. a. Patient with a supranasal and anterior orbital encephalocoele. Coronal incision, frontal craniotomy, bone graft to defect and medial canthoplasty. b. 6-year postoperative view.

Discussion

Encephalocoeles are relatively rare deformities, and even busy craniofacial units may only see a handful of these cases. In addition, there is a vast array of presentations of encephalocoeles. Therefore, it makes it difficult to propose management strategies that are based on good clinical trials and scientific evidence. In such rare deformities, it may be useful to have a classification system that has clinicopathological relevance. It is akin to the classification of rare facial clefts proposed by Tessier. This would enable better evaluation of procedures and outcomes and have a language and communication that is understood by those in that particular field. Whilst the Suwanela classification is the most comprehensive thus far, there was no obvious clinicopathological correlation. In addition, as these were all post-mortem specimens, there were no outcomes based on surgery.

We have proposed a classification based on these cases which has clinicopathological relevance. Present-day imaging allows us to more accurately appreciate anatomical changes and pathways of the encephalocoele. This has made it possible for us to plan surgery more accurately and help us achieve a better outcome. It is a more complete and comprehensive classification that includes patients that have encephalocoeles that were once regarded as ‘theoretical’. This classification has the potential to accommodate newer types of encephalocoeles that are periodically described as case reports.

The occipital and cranial vault groups of Suwanela are grouped as one ‘cranial’ in our proposal. We divide the Suwanela frontoethmoidal group into ‘nasal’ and ‘orbital’. We have termed their cranioschisis group ‘facial cleft’.

The surgical correction of encephalocoeles is a paper on its own. However, there are certain principles we employ. Firstly, we try to minimize the visible scarring. Therefore, a coronal approach is usually used. We try to avoid any incisions on the dorsum of the nose as these are usually apparent. In the nasal group, if there is mild to moderate excess skin over the nasal dorsum, it has the ability to contract with time. If there is a major excess, we use a paranasal incision as these scars are

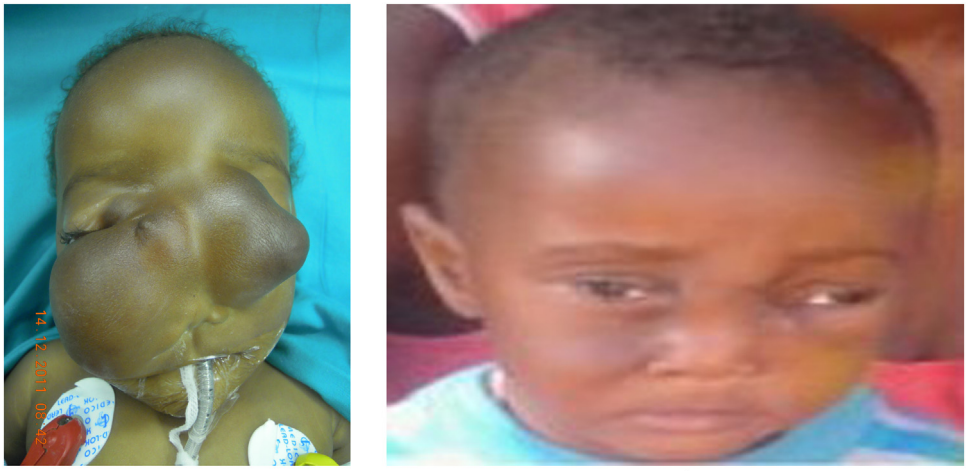

Fig. 9. a. Patient aged 10 months with infranasal encephalocoele. Coronal and bilateral paranasal incisions, bone graft to defects, medial canthoplasty. b. 1-year postoperative view.

camouflaged in a naturally hidden area. If the surgical approach can be performed without a frontal craniotomy, this extracranial method is preferred. Good dural closure and tissue glue are used to obtain a watertight closure. The bony defect is closed with a cranial bone graft. A dorsal nasal bone graft and a medial canthoplasty are performed if required.

This classification system will allow better communication amongst craniofacial teams. It will also afford us the chance to analyse outcomes and procedures in similar groups of patients. Modifications in strategies, staging, timing and technical surgical details will ensure to produce superior clinical outcomes.

Conclusion

A classification of encephalocoeles is proposed that is based on the second largest series to date. This is an updated version of previous classifications that is easy to understand, has clinicopathological relevance, incorporates newly described encephalocoeles and should allow better communication amongst those involved in the care of these patients.

Declaration of Competing Interest

None declared.

Funding

None declared.

Ethics Approval

Ethical approval was obtained from the University of Kwazulu Natal. BREC/00001834/2020. Patient consent was obtained for publishing photographs.

References

1. Suwanwela C, Suwanwela N. Air study in infants and children with frontoethmoidal encephalomeningocele. *Neuroradiology*. 1972;4(3):190–194.

2. David DJ. Cephaloceles: classification, pathology, and management–a review. *The Journal of craniofacial surgery*. 1993;4(4):192–202.
3. Rojvachiranonda N, David DJ, Moore MH, Cole J. Frontoethmoidal Encephalomeningocele: New Morphological Findings and a New Classification. *Journal of Craniofacial Surgery*. 2003;14(6):847–858.
4. Mahapatra A. Anterior encephalocele - AIIMS experience a series of 133 patients 2011. S2730 p.
5. Mahatumarat C, Rojvachiranonda N, Taecholarn C. Frontoethmoidal encephalomeningocele: surgical correction by the Chula technique. *Plast Reconstr Surg*. 2003;111(2):556–565 discussion 667.
6. Arifin M, Suryaningtyas W, Bajamal AH. Frontoethmoidal encephalocele: clinical presentation, diagnosis, treatment, and complications in 400 cases. *Child's nervous system: ChNS: official journal of the International Society for Pediatric Neurosurgery*. 2018;34(6):1161–1168.
7. Shokunbi T, Adeloye A, Olumide A. Occipital encephalocoeles in 57 Nigerian children: a retrospective analysis. 1990. 99-102 p.
8. Albano L, Parisi V, Acerno S, Barzaghi LR, Losa M, Mortini P. Sublabial transsphenoidal microsurgical technique to treat congenital transsphenoidal encephalocele: a technical note. *Neurosurgical review*. 2019;42(2):571–575.
9. Govindaraju R, Tang IP, Prepageran N. Management of sphenoid lateral recess encephalocoeles. *Curr Opin Otolaryngol Head Neck Surg*. 2019;27(1):37–46.
10. Moore MH, Lodge ML, David DJ. Basal encephalocoele: imaging and exposing the hernia. *Br J Plast Surg*. 1993;46(6):497–502.
11. Nagata Y, Takeuchi K, Kato M, Chu J, Wakabayashi T. Lateral temporal encephaloceles: casebased review. *Child's nervous system: ChNS: official journal of the International Society for Pediatric Neurosurgery*. 2016;32(6):1025–1031.
12. Sdano MT, Pensak ML. Temporal bone encephaloceles. *Curr Opin Otolaryngol Head Neck Surg*. 2005;13(5):287–289.
13. Tuncbilek G, Calis M, Akalan N. Spontaneous lateral temporal encephalocele. *The Journal of craniofacial surgery*. 2013;24(1):e90–e92.
14. Tyler-Kabara EC. Sphenoidal encephaloceles. *Journal of neurosurgery Pediatrics*. 2013;11(5):504 discussion.
15. Worrall DM, Pai A, Garneau JC, Perez ER, Cosetti MK, Smouha EE, et al. Temporal Bone Encephaloceles: Utility of Preoperative Imaging. *Otolaryngol Head Neck Surg*. 2020;163(3):577–581.
16. Asadi H, Morokoff A, Gaillard F. Occult temporal lobe encephalocoele into the transverse sinus [In Process Citation] 2015. 1202-4 p.
17. Moore M, Lodge M, David D. Basal encephalocoele: imaging and exposing the hernia1993. 497-502 p.
18. Asadi H, Morokoff A, Gaillard F. Occult temporal lobe encephalocoele into the transverse sinus. *J Clin Neurosci*. 2015;22(7):1202–1204.
19. Benson JC, Lane J, Geske JR, Gompel JV, Krecke KN. Prevalence of Asymptomatic Middle Cranial Fossa Floor Pits and Encephaloceles on MR Imaging. *AJNR Am J Neuroradiol*. 2019;40(12):2090–2093.
20. Sare G, Varma A, Green K, Herwadkar A, Gnanalingham K. Pneumococcal meningitis secondary to intra-sphenoidal encephalocoele2009. 103-4 p.
21. Jain A, Tullu MS, Agrawal M, Jadhav DU. Occult Encephalocele Causing Recurrent Meningitis. *Pediatr Neurol*. 2015;53(3):270–271.
22. Sare GM, Varma A, Green K, Herwadkar A, Gnanalingham KK. Pneumococcal meningitis secondary to intra-sphenoidal encephalocoele. *Acta neurochirurgica*. 2009;151(1):103–104.
23. Sheikh SI, Vaitkevicius H, Mez J, Diamond EL. Meningitis associated with sphenoid sinus encephalocoele. *Pract Neurol*. 2010;10(1):43–44.
24. Wan DC, Lazareff JA, Jarrahy R, Bradley JP. Correction of large facial encephalocele with bilateral rare craniofacial clefts. *The Journal of craniofacial surgery*. 2011;22(1):338–342.
25. Hunt JA, Hobar PC. Common craniofacial anomalies: facial clefts and encephaloceles. *Plast Reconstr Surg*. 2003;112(2):606–615 quiz 16,722.