Nasal Encephalocele: Endoscopic excision with anesthetic consideration

Introduction
An encephalocele is a herniation of cranial contents through a defect in the skull. It may include meninges only, termed as meningocele, or it may contain brain tissue and meninges, termed as meningoencephalocele. An encephalocele that has lost their intracranial connection is termed as a glioma; 15% of them remain connected to the central nervous system (CNS) via a fibrous stalk [1]. The first medical report of encephalocele may have appeared in the 16th century [2]. This condition occurs in 1 of every 4000 live births and is equally distributed between males and females, the majority of encephaloceles are seen in early childhood [3]. Encephaloceles may present as nasal broadening and/or as a blue, pulsatile, compressible mass near the nasal bridge which transilluminates, enlarges with crying or with bilateral compression of the internal jugular veins (Furstenberg test), or as an intranasal mass arising from the cribriform plate [4,5].
An intranasal encephalocele presents with a mass in the nose; it may be mistaken for a nasal polyp and removed with disastrous complications [6]. The presence of intracranial communication makes biopsy contraindicated, also rough manipulation of the mass should be avoided because of the risk of meningitis and because compression may cause somnolence or even seizures in some cases [1]. Children with encephalocele may have hydrocephalus and increased intracranial pressure (ICP) [7], lowering of the CSF pressure intra- and post-operatively is usually needed in order to decrease the CSF leak and to improve the surgical outcome. Estimation of fluid balance in young children may be difficult particularly after mannitol therapy or blood loss [8]. Such anesthetic problems need close observation.
The aim of this study was to evaluate the efficacy of endoscopic removal of intranasal encephalocele, also to document the role of anesthetist in the operative and postoperative periods.

Methods
This study was conducted on 9 patients that had been diagnosed to have nasal encephalocele. The cases were admitted and treated in the Hospitals of Cairo University; Department of Neurosurgery (5 cases), Department of Otolaryngology (2 case), and the Pediatric unit of Otolaryngology (2 cases) in the period from December 2002 to January 2008. Five males and four females, their ages ranged from 8 months to 3 years and 9 months (with a mean age of 21 months). After discussion with the parents and obtaining written consents; surgical removal of the masses was carried out for all cases. Follow up was carried out for at least 21 months.
The following management protocol was applied for all cases

- Preoperative evaluation:
  . Complete history taking and clinical examination
. Computerized tomography (CT) and/or magnetic resonance imaging (MRI).
. Complete blood picture as a routine preoperative preparation.
. Preoperative medication: all patients were fasting for at least 2 hours before the procedure, and they arrived in the operating room with an intravenous cannula in situ.

All children were premedicated -while still being in the ward with their parents in a room outside the cath lab- with IV dose of midazolam (*Dormicum*® *Roche Hoffmann-La Roche Ltd.Basel, Switzerland*) 0.1mg/kg over a 5-minute period, given by the attending anesthesiologist just before the procedure, and then the anesthesiologist accompanied the child to the operating room.

- **Intervention:**
  On arrival the operating room, and prior to induction of anesthesia, all patients were connected to standard monitors that included five leads electrocardiogram(ECG), and ECG leads II and V5 were continuously monitored, a noninvasive arterial pressure (*Dinamap, Criticon, CA, USA*), and a digital pulse oxymetry (*Novametrix,515C, NY, USA*). Heart rate (HR), digital oxygen saturation (SpO2), and mean arterial blood pressure (MAP) were recorded every 5 min. Heat loss should be minimized; therefore the child should lie on a heated mattress and covered with warming blankets. All patients received atropine 0.01 mg/kg IV.

Inhalational induction of anestheisia was performed using sevoflurane, and ventilation was controlled as soon as possible after inhalation induction, and the concentration of inhalation agent decreased. Sevoflurane is used as its effects on CSF pressure are minimal [9] and induction is rapid and smooth.

Oral intubation and insertion of temperature probe, and oropharyngeal pack is mandatory in cases of endoscopic excision of nasal encephalocele, nasal intubation is inappropriate as it narrows the surgical field and may cause trauma to the sac.

Following securing of the airway with an endotracheal tube, a second IV line was obtained. Intravenous access should be obtained with a large-bore cannula as is reasonable based on the patient’s size. Note that the saphenous vein is usually quite large and a 22 g or larger cannula can usually be placed for later volume resuscitation even in infants [10]. If not previously obtained, a sample should be sent to the blood bank for cross-match of two ‘adult’ units of packed red cells. At the time of incision, mannitol (1 g/kg) is administered. A brisk diuresis usually ensues shortly after administration and it is important to closely monitor hemodynamic parameters to maintain adequate intravascular volume. Once the surgery is started, furosemide (0.5–1 mg/kg) may be administered if the sac looks ‘tight’, but this is usually not required after mannitol administration [11]. Ventriculo-peritoneal shunt was done for cases presented with hydorccephalus before starting the nasal procedure. Endoscopic excision of the nasal mass was done after identification of their stalk that connects it to the intracranial structure, cutting of the stalk was carried out with the help of bipolar diathermy. The bony defect was palpated and packed with fat that taken from the thigh during harvesting fascia lata graft, the area round the defect was refreshed then the
defect was sealed with the graft (in an overlay fashion) and supported with gel foam (Gelfoam; Pfizer Inc, New York, NY). The operated side of the nose was packed for 2 days using sterile gauze.

Fluid requirements are the same as any other paediatric case, based on weight of the baby. Preoperative dehydration is common in children with intracranial hypertension. Isotonic crystalloid is the intravenous fluid of choice for intraoperative maintenance and hydration. Hypotonic fluid, e.g. Ringer’s lactate (273 mOsm/L) can exacerbate brain edema. Glucose containing fluids may increase the risk of neurologic injury. Intraoperative blood glucose assessment is recommended with administration of exogenous glucose on an as needed basis only, except in patients at high risk of hypoglycemia (e.g. neonates) [12]. Continuous assessment of blood loss is required. Some patients will require replacement with red blood cells. It is best to wait until the hemoglobin drops to about 8 g/dl before transfusing unless the infant has heart disease or other co-existing pathology requiring a greater oxygen carrying capacity. At that point, a continuous transfusion totaling 20–25 ml/kg over the remaining course of the procedure will usually increase the hemoglobin to 12–14 g/dl by the end of the procedure [10].

At the end of the surgical procedure, the patient can usually be safely extubated, placed in a head up position, and transported on oxygen to the intensive care unit. Deep extubation may carry the risk of respiratory obstruction or aspiration, so awake extubation is preferred provided that the patient is in good analgesia to avoid any stress that may result in increased ICP.

All surgical specimens were sent to the pathology department for histopathological examination except of one case in which the sac contained fluid only with no soft tissue mass.

- Postoperative care and follow up:

Careful assessment of the airway must be done during the early postoperative period. The patient is also observed for signs of irritability, nausea or vomiting, or decreased neurological function which might be indicative of increased ICP. The patient is maintained in a horizontal position, either prone or lateral for several days until the dural closure has had an opportunity to begin healing. This decreases the risk of CSF leak [13].

All patients were admitted for one week post-operatively and they were received antibiotic injection through this period with removal of nasal pack after the second postoperative night. Continued care to maintain a normal or even decreased CSF pressure is essential, and in this study this was maintained using furosemide (0.5–1 mg/kg) every 8 hours for the 2 days postoperatively. Nasal endoscopy was done for all patients 3 months postoperatively and then yearly and on occurrence of nasal symptoms such as suspicious of CSF leak; the patients were sedated using IV midazolam in a dose of 0.1mg/kg over a 5-minute period just before the procedure.

Results
Nine children were included in this study; the parents of all children gave no history of similar conditions in the family. No cases gave history of meningitis or previous surgical intervention for their lesions. Three cases presented with CSF rhinorrhea in addition to nasal obstruction, while 4 cases presented with unilateral nasal obstruction only. Two patients presented with enlarged head circumference and associated hydrocephalus with no nasal symptoms. A swelling was detected at the right medial canthus in one case, while midfacial cleft was detected in another case, three cases showed hypertelorism. None of the patients had neurological deficit or evidence of mental retardation. On endoscopic examination, all cases showed unilateral nasal polypoid mass that enlarge on crying and on compression of the jugular vein (Furstenberg test). Radiological study (CT for 3 cases, MRI for 4 cases, and CT and MRI for 2 cases) showed unilateral nasal mass with skull base defect in the area of the cribriform plate in eight cases (Fig 1), while one case showed unilateral ethmoidal mass with the orbit pushed laterally and a bony defect in the fovea ethmoidalis (Fig 2). According to the modified classification of Rosenfeld and Watters [14], eight of our cases had basal intranasal encephalocele while one case had sincipital nasoethmoidal lesion.

Regarding the associated congenital anomalies, 1 case presented with facial cleft and hypertelorism (Fig 3), 2 cases presented with hypertelorism, and 6 cases showed no anomalies. Intra-operatively CSF leakage was not a problem and none of our cases showed complications. The skull base defect was seen in the cribriform plate in eight cases, while one case had ethmoidal roof defect that necessitated ethmoidectomy for good exposure and closure of the defect. Endoscopic follow-up showed closure of the skull base defect with no CSF leak.

Discussion
Encephaloceles are extracranial herniations of the meninges and/or brain which maintain a subarachnoid connection. The lesions are classified as occipital, sincipital, and basal, based on the location of the skull base defect [14,15]. Occipital are the most common representing 75%. Sincipital are frontonasal lesions which present as a mass over the nose, glabella, or forehead. The intracranial connection is usually anterior to the cribriform plate [16]. Suwanwela and Suwanwela [17] divided nasal encephaloceles into nasofrontal, nasoethmoidal, and naso-orbital lesions based on the projection of the mass between the nasal and frontal bones, along the side of the nose, or into the medial orbit. Basal lesions make up about 10% of lesions and present as an intranasal or nasopharyngeal mass. Basal lesions herniate either through the cribriform plate or posterior to it which explains their presentation in the nose instead of externally [16].

The embryologic development of encephaloceles may be due to failure of the fonticulus frontalis -space between frontal and nasal bones- to close properly which lead to a herniation of intracranial contents that maintains its connection to the subarachnoid space [1].
The disease should be differentiated from other pediatric midline nasal masses; dermoid sinus cysts present as a mass on the dorsum of the nose or intranasally, with a pit or sinus tract opening on the nasal dorsum, hair around the external opening, and discharge of pus or sebaceous material. Nasal gliomas are firm masses which are non-pulsatile, present on the nasal dorsum and/or arise from the lateral nasal wall, have telangiectasias of the overlying skin, and do not enlarge with bilateral compression of the internal jugular veins (Furstenberg test). Encephaloceles may present as nasal broadening and/or as a blue, pulsatile, compressible mass near the nasal bridge which transilluminates, enlarges with crying or with bilateral compression of the internal jugular veins, or as an intranasal mass arising from the cribiform plate. When a child is presented with a unilateral nasal mass, a high index of suspicion is required. The diagnosis is confirmed by CT and/or MRI imaging. Image findings include soft tissue mass, fluid filled cyst, intracranial mass, enlargement of the foramen cecum, and distortion of the crista galli. CT imaging better delineates bony abnormalities while MRI is valuable to identify an intracranial connection. Due to increased cost of two tests, delay in diagnosis, and added risk of additional anesthesia for additional imaging; some authors recommend MRI as the initial imaging study. Schlosser et al. used preoperative three dimensional CT scanning in the diagnosis of encephalocele, they found it useful to demonstrate the full extent of the skull base defect and to provide images to use in counseling of the parents more easily understood than conventional two dimensional CT. In our study, five of our cases have already undergone CT before presentation; in two of them the intracranial connection was not apparent and MRI was requested, while we requested MRI for the last four cases on initial presentation which was enough in demonstrating the lesions and their intracranial connections. The incidence of hydrocephalus in patients with encephaloceles is reported to be about 50%. Shilpakar and Sharma reported hydrocephalus in 6 (37.5%) out of their 16 cases, 3 of them needed CSF diversion. In our study, we found 2 (22.2%) out of 9 cases had associated hydrocephalus, and they were treated with ventriculo-atrial shunt. One of our cases presented with midfacial cleft, an association that had been reported before by Kawakami et al, Wexler et al and Sharif et al, however some authors considered basal encephalocele is a part of median cleft face syndrome. Hypertolerism was detected in 3 (33.3%) of our cases, a finding that was detected by many authors in cases with basal encephalocele. The proper management of nasal encephalocele requires a multidisciplinary approach that includes otolaryngology, neurosurgery, neuroradiology, and pediatric anesthesia services. Patient should undergo a complete examination to exclude any other congenital abnormalities. Nasal endoscopy is performed to determine the location, origin, and extent of the nasal mass and to assess lack of or presence of pulsation, which is of paramount importance. After thorough evaluation, definitive treatment is surgical. Most authors agree that encephalocele should be managed early in life, this makes identification of the intracranial connection technically easier and allows more complete repair of the
dural defect. When removal is indicated, there are multiple surgical approaches; including lateral rhinotomy, a transnasal approach, a coronal flap approach according to the location of the lesion [1]. However, the treatment of a basal intranasal encephalocele using transnasal endoscopic approach could obviates the possible morbidity associated with transcranial approach such as loss of sense of smell, post-operative intracerebral hemorrhage, cerebral edema, epilepsy, frontal lobe dysfunction with memory and concentration deficits, in addition to avoiding the postoperative scar that may follow approaches other than endoscopic approach [6, 26]. All cases of our study were treated transnasally, endoscopic approach was efficient in exposure and removal of the mass. The skull base defect was closed endoscopically using fascia lata, we achieved a success rate of 100% with no morbidity or mortality. Endoscopic removal of intranasal encephalocele had been reported by many authors [3, 27,28,29]. Woodworth et al [27] treated eight cases of congenital CSF leak and encephalocele via endoscopic approach; they used temporalis fascia in closure of the skull base defect in most cases, while cases with large skull base defects needed composite turbinate graft and hydroxyapatite or sometimes mastoid cortical bone graft, their treatment was successful on first attempt with no morbidity except in one case that experienced nasal stenosis postoperatively. Nogueira et al [28] treated a 2-year-old girl with a meningo-encephalocele after episodes of meningitis; they used transnasal endoscopic approach and they closed the skull base defect with nasal septal flap pedicled on the sphenopalatine artery. Rahbar et al [3] used a combined frontal craniotomy and transnasal endoscopic approach in one of their six encephaloceles with no recurrence for one year. However, continuing progress in the surgical management of congenital skull base defects demonstrates that endoscopic repair is a successful alternative to traditional craniotomy approaches, with less morbidity [27]. The use of premedication in those children should be considered to facilitate smooth induction and decrease agitation which may lead to elevation of the ICP due to crying and breathholding. According to Feld et al [30], Midazolam was used in our study to achieve this goal. However, over-sedation of infants should be avoided as it may result in decreased respiratory effort and increases in ICP secondary to hypercarbia. Also, maintenance a normal or even a low CSF pressure is important intra-operatively and in the postoperative period to avoid leakage of CSF through the wound. All these points call for co-operation between surgeons and anesthetists to obtain the best operative outcome.

**Conclusion**

Endoscopic excision of intranasal encephalocele is an effective method with high success rate and low morbidity. Anesthetist plays an important role in the operative and postoperative period, even during the endoscopic follow up; sedation of the children is usually needed.

**References**


**Figure Legends**

Fig 1 Radiological evaluation of a patient with basal encephalocele, CT and MRI show left nasal mass with a defect in the cribriform plate.

Fig 2 Radiological evaluation of a patient with nasoethmoidal encephalocele, CT shows a defect in the fovea ethmoidalis (arrow head) and MRI shows right ethmoidal mass (arrow).

Fig 3 Patient with midfacial cleft (after repair of his cleft lip and palate), his CT shows a defect in the cribriform plate with a mass in the left nasal cavity.