Child's Nervous System (2019) 35:2091–2098
https://doi.org/10.1007/s00381-019-04167-3

FOCUS SESSION

Endonasal endoscopic skullbase surgery in children

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Received: 11 April 2019 / Accepted: 17 April 2019 / Published online: 12 May 2019

Abstract

Objective The role of endonasal endoscopic approach for pathologies in the paediatric population is evolving and has still not been accepted as standard of care in neurosurgery. It represents a challenge in terms of narrow access, instrument manipulation and adequate reconstruction of defects. We have described our experience in 49 cases from a single neurosurgical unit in paediatric skull base surgeries through this approach over the last 12 years.

Material and methods A case series of 59 paediatric skull base surgeries in 49 children through endoscopic endonasal route over the last 12 years is presented. The age ranges from 4 months to 18 years. Out of 49 cases, 22 cases were of craniopharyngiomas, 8 cases of pituitary adenomas, 5 cases with CSF rhinorrhea, 5 cases with meningoencephalocele, 3 cases of Rathke's cleft cysts, 2 cases of odontoidectomy and 4 miscellaneous cases viz. mucocele, hypothalamic glioma, esthesioneuroblastoma and epidermoid. CSF leaks were repaired with free graft in the initial years and by vascularized flap more recently.

Results The goal of surgery was achieved in all but two cases in whom the tumour excision was unsatisfactory due to failure of the cyst wall to collapse after decompression. Extent of tumour excision was not compromised by the choice of this approach. Revision surgery for CSF leak was required in three patients. Local vascularized nasoseptal flap has been possible even in very young patients and has now become the standard for reconstruction.

Conclusion In spite of the challenges posed by small nostrils and ill-developed sinuses in the paediatric age group, surgery from endoscopic endonasal corridor is possible to be carried out successfully in selected cases.

Keywords Endonasal · Endoscopy · Paediatric · Skullbase

Introduction

Paediatric skull base lesions are infrequently encountered and their management poses several challenges. Progress in skull base surgery over the last three decades has mainly addressed the adult skull base tumours. Endonasal endoscopic approaches have added a new dimension to these surgical approaches with their success through minimal invasion, and the concept of teamwork in surgical management of skull base tumours has further evolved. Several congenital as well as other paediatric skull base lesions may also require these approaches in, and a combination of various approaches may occasionally demand teamwork of neurosurgeons, ENT surgeons and craniofacial plastic surgeons.

The endonasal endoscopic approaches in young patients have certain limitations because of small nares, developing air sinuses and possible interference with craniofacial growth. Repair of the defects at the skull base at this age is also believed to have limited options and require innovative solutions.

We present our experience with 49 children with midline skull base lesions primarily treated with endonasal endoscopic surgery over the last 12 years. Goal of the surgery was achieved in all but two cases. Skull base repair with local vascularised flaps was performed in most cases in recent years with a remarkably successful outcome.

Materials and methods

A case series of paediatric skull base surgeries through endoscopic endonasal route over the last 12 years is

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presented. The age ranged from 4 months to 18 years. Thirty-one children were less than 12 years of age, and eighteen children were of the 13 to 18 years age group (Table 1). Preoperative assessment included MRI brain, CT paranasal sinuses, visual perimetry, serum hormones and baseline electrolyte levels. Out of 49 cases (Table 2), 22 cases were of craniopharyngiomas, 8 cases of pituitary adenomas, 5 cases of CSF rhinorrhea, 5 cases of meningoencephalocele, 3 cases of Rathke's cleft cysts, 2 cases of odontoidectomy and 4 miscellaneous cases comprising of hypothalamic glioma, epidermoid, mucocele drainage and esthesioneuroblastoma biopsy. A total of 59 endonasal procedures were performed on these patients (Table 3). Skullbase exposure was tailored to the pathology being dealt with. The floor of the sella was drilled with a number 3 or a number 4 rough diamond drill with paintbrush-like swipes till the four blue lines were seen (viz. 2 intercavernous sinus above and below and the 2 cavernous sinus laterally). The last layer was peeled away using a dissector. Standard dural incision and tumour excision techniques were applied. CSF leaks were repaired with free graft in the initial years and by vascularized Hadad Bassagasteguy flap from 2009. Local vascularized nasoseptal flap was possible to obtain even in very young patients and has now become our standard of care for reconstruction of skullbase. For tumours with parasellar or suprasellar extension, intraoperative neuronavigation and Doppler was used as an adjunct to achieve maximal safe resection. Each patient underwent two sessions of nasal cleaning and assessment of reconstructed skullbase at postoperative day 3 and postoperative day 7 respectively. Lumbar drain was kept for 72 hours in those patients who developed CSF leak intraoperatively. Serum electrolytes were checked routinely for every patient on the evening of surgery and 12 hourly for 3 days till the patient was discharged. Patients who underwent planned subtotal resection or those with a residual lesion on follow-up MRI after 10 weeks were advised adjuvant radiation therapy.

Table 1 Age group distribution of the cases

Diagnosis	Number
Craniopharyngioma	22
Pituitary adenoma	8
CSF leak repair	5
Meningoencephalocele	5
Rathkes cleft cyst	3
Odontoidectomy	2
Miscellaneous (mucocele, hypothalamic glioma, esthesioneuroblastoma, epidermoid)	4
Total	49

Table 2 Distribution of cases as per diagnosis

Age group	Number
0–4 years	10
5–12 years	21
13–18 years	18

Results

The goal of surgery was achieved in 47 out of 49 cases. Tumour was not satisfactorily excised in two children due to the failure of cyst wall to collapse during surgery. One child underwent repeat endonasal surgery before discharge while the other child underwent a transcranial reservoir insertion into the cyst wall. Both the cases have been advised adjuvant radiation therapy. Extent of tumour excision was however, not compromised by the choice of this approach.

Amongst the five cases with meningoencephalocoele, four cases were of basisphenoidal encephalocele repair and one case of meningocele repair. Combined transcranial with endonasal repair was required for three out of the four encephalocele cases (Fig. 1). A 7-month child eventually required a craniofacial reconstruction of the nasal bridge and correction of hypertelorism after 4 years of combined encephalocele repair.

Non aeration of sphenoid sinus was not a limiting factor, though it did represent an important technical consideration preoperatively (Fig. 2). This was overcome by availability of better drilling instruments, use of navigation and doppler guidance and an endoscope providing a panoramic view.

Eight cases of pituitary adenomas were operated in this series. This comprised three cases of Cushings disease (Fig. 3), one case of gigantism and four non-secretory cases presenting with either apoplexy or visual dysfunction. Although satisfactory excision was achieved in all these cases, one child with Cushing's disease did not achieve satisfactory fall in adrenocorticotrophic hormone (ACTH) levels and was subjected to adjuvant radiation therapy.

Extended endonasal approach was applied for cases with Rathke's cleft cyst (Fig. 4) and craniopharyngiomas (Figs. 5, and 6).

Two cases of congenital cranio-vertebral junction anomaly underwent transoral with transnasal odontoidectomy (Fig. 7).

Table 3 Surgical procedures performed

Procedure	Number
Primary endonasal surgery	49
Redo endonasal tumour excision	6
Redo endonasal CSF leak repair	4
Total	59

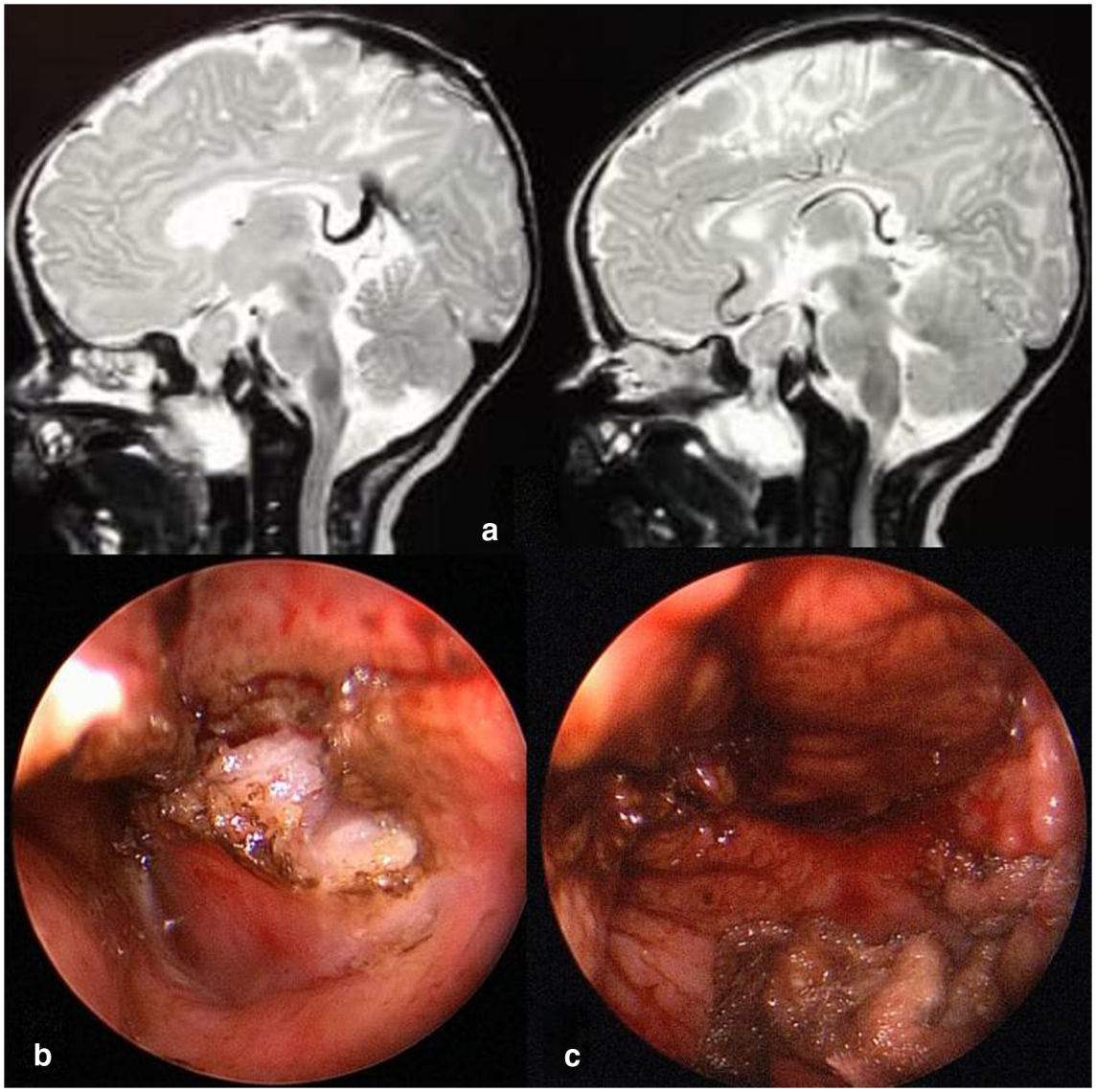

Fig. 1 **a** MRI brain sagittal view T2-weighted image of a 7-month old girl showing a basi-sphenoidal encephalocele. **b** Intraoperative picture of the encephalocele with the defect. **c** Intraoperative picture of the vascularized naso-septal flap placed for repair of the skullbase defect. This child underwent a combined transcranial repair of the frontal encephalocele component. She required a craniofacial reconstruction of the nasal bridge and correction of hypertelorism after 4 years

Two patients with craniopharyngiomas and two with pituitary adenomas developed transient diabetes insipidus which resolved by the time they were discharged.

CSF leak was present in four cases intraoperatively which was managed conservatively with lumbar drain. Amongst them was a child of craniopharyngioma with flare up of a preexisting Stevens Johnsons syndrome. Revision surgery for CSF leak was required in three patients (6.12%). Ten children were subjected to repeat surgeries (6 endonasal and 4 transcranial) either due to recurrence (8/10) or due to unsatisfactory removal of tumour in primary surgery (2/10).

There was one surgical procedure-related death in this series of a child aged 5 years with craniopharyngioma who expired on postoperative day 7 due to ventriculitis.

Discussion

The earliest reports focussing on the use of endoscopic endonasal skull base surgery in children for congenital lesions were published in plastic, reconstructive, head and neck surgery literature [1, 2] and for nasopharyngeal tumours in ENT literature [3].

The main anatomical consideration in this surgery relates to the development and pneumatization of the paranasal sinuses. Maxillary sinus is the first to develop in children and reaches adult size by 18 months. 45.7% are pneumatized by the first month of life and over 97% by the age of 1 year. The ethmoidal sinuses are fluid filled at birth and pneumatized completely by the age of 12. Sphenoid sinus is identifiable as early as 2 months

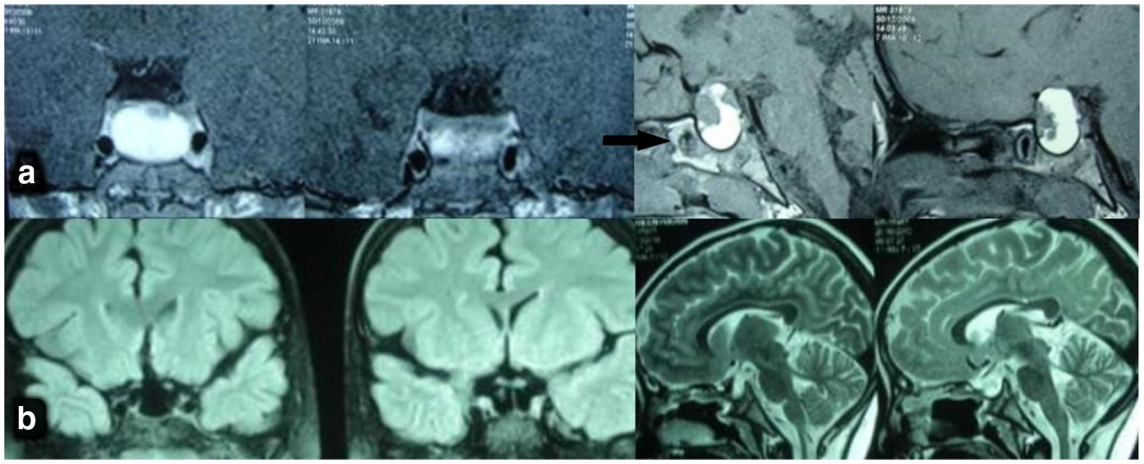

Fig. 2 **a** MRI brain coronal and sagittal views showing non-pneumatization of the sphenoid sinus (black arrow) in a 7-year old boy with craniopharyngioma. **b** Postoperative MRI brain coronal and sagittal images showing complete excision of the lesion. Navigation-guided drilling of the sphenoid sinus was done till the four blue lines were seen along with the optico-carotid protuberances

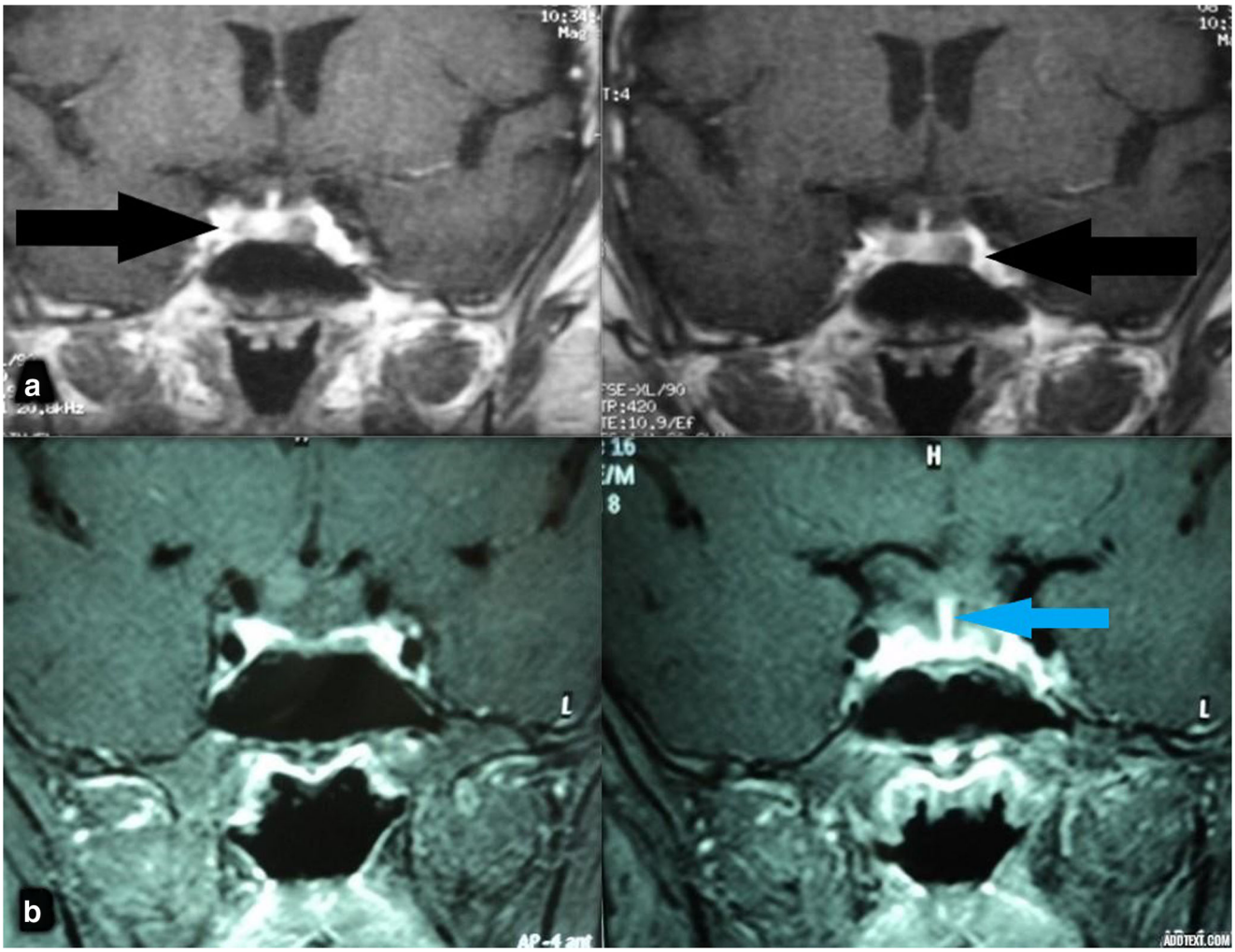

Fig. 3 **a** MRI brain coronal view of a 15-year-old boy with Cushings disease showing a rare occurrence of double pituitary microadenoma (black arrow). **b** Postoperative MRI brain coronal view showing complete excision of the adenoma with intact pituitary tissue and infundibular stalk (blue arrow). The patient has been in remission for 5 years now and requires no hormonal supplements

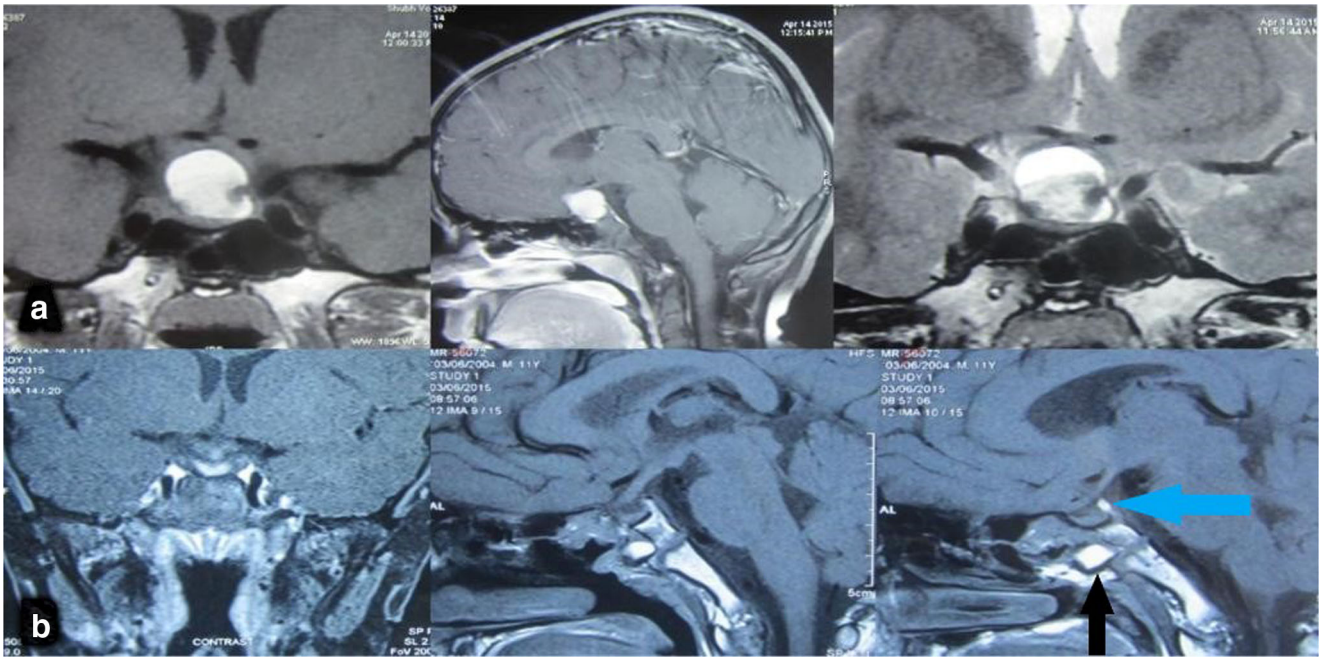

Fig. 4 **a**) MRI brain images of a 12-year old boy with Rathke's cleft cyst. **b** Postoperative T1-weighted MR images of the boy showing complete excision of the cyst. Fat and naso-septal flap used for repair of the defect (black arrow) and posterior pituitary (blue arrow) is seen. Patient developed CSF leak and underwent exploration for the same after 12 days which showed displacement of the flap. This was repositioned and reinforced with glue

of age and in 100% of subjects at the age of 3 years on axial image and by 6 years on sagittal image. The sinus reaches its full size by late teenage years. Frontal sinus is the last to pneumatize. Only 8% are pneumatized up to 2 years while 97% show pneumatization by 15 years of age [4]. A radio anatomic study has shown that the piriform aperture width is

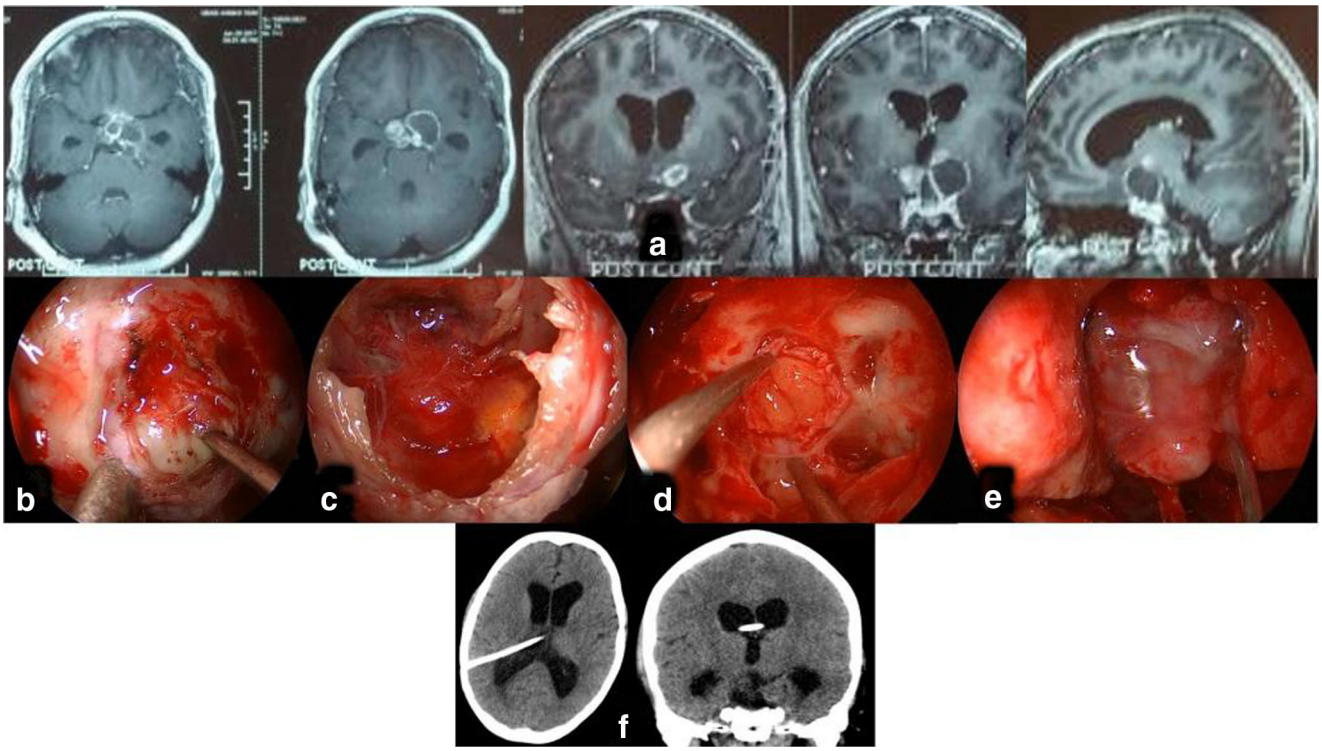

Fig. 5 **a**) MRI brain (post contrast) views of a 16-year old boy showing a sellar suprasellar solid cystic craniopharyngioma. **b** Intraoperative picture of the tumour excised via endonasal route. **c** Tumour bed after a planned subtotal excision of the tumour. **d** Fat placed in the defect with inlay fascia lata graft. **e** Fibrin glue applied over the fascia and fat. **f** Postoperative CT scan of the child

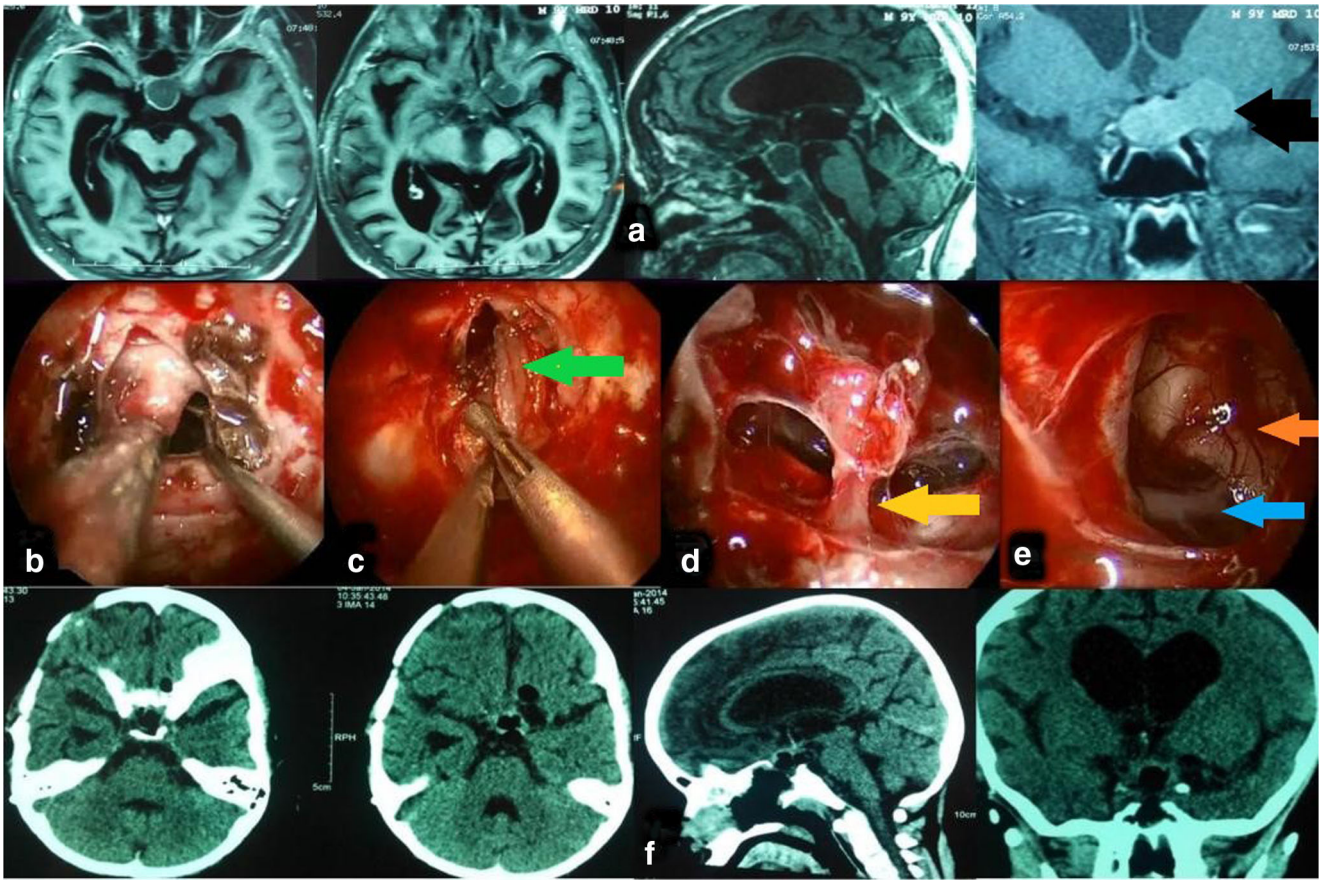

Fig. 6 **a** MRI images of an 8-year old boy with craniopharyngioma with a large left parasellar extension. **b** Intraoperative picture of the tumour excised via endonasal route. **c** The parasellar extension (green arrow) was completely excised via suprasellar approach. Intraoperative navigation and Doppler assistance was taken. **d** The intact pituitary stalk (yellow arrow) seen after gross total resection of the tumour. **e** Carotid artery (blue arrow) and oculomotor nerve (orange arrow) seen after resection of the tumour. **f** The boy developed transient diabetes insipidus which resolved prior to discharge. Postoperative CT scan of the child showing completeness of the excision

significantly greater beyond the age of 7 years. Three fourth of the anterior sellar floor is pneumatized by 7 years while posterior pneumatization on the clivus is seen only after 12 years of age. Internal carotid distance does not show much changes in different age groups [5]. A detailed radiological study by Weil Cornell group [6] has found that a wider inter-carotid distance at the superior clivus (mean 20.75 mm) and shorter nare-dens working distance predict a better outcome in paediatric patients undergoing this approach.

Although our series had one patient who required repair of the craniofacial defect after 5 years of endonasal surgery, it has been found that the long-term impact on midface craniometrics in paediatric population after endonasal surgery does not differ as compared to older age group patients [7].

The indications for endonasal endoscopic surgery in children include cerebrospinal fluid (CSF) leak repairs, development anomalies such as meningoencephalocoele and odontoid invagination. In teenagers, more common indications are pituitary tumours, craniopharyngiomas and occasionally chordomas.

The challenges in paediatric skull base surgeries include considerable blood loss related to exposure. Several anatomical issues need to be tackled in endonasal surgery viz. narrow corridor due to small nasal apertures, ill-defined turbinates, delayed development of paranasal sinuses and narrow inter-carotid distance. Paediatric endoscope should be used without any sheath and cleaning device. Lack of dedicated instrument set also can be challenging. We have used micro ear instrument set for most of the nasal manoeuvres especially for children less than 4 years. The worry about improper growth of the face also restricts options for widening the approach and subsequently of reconstruction.

Clinical experience with six patients below 12 years of age [8] for repair of paediatric meningoencephalocoele has shown good results. There have been several reports of pituitary tumours in children being treated successfully in children by transsphenoidal approach [9, 10]. Most of them are functioning tumours and need surgery, except those with prolactinomas. More recently, a similar success has been reported by endonasal endoscopic approach as well [11].

Surgical resection of paediatric craniopharyngioma has been advocated by Laws et al. [12]. Patel et al. [13] from the Stanford group were able to achieve higher rates of total resection with

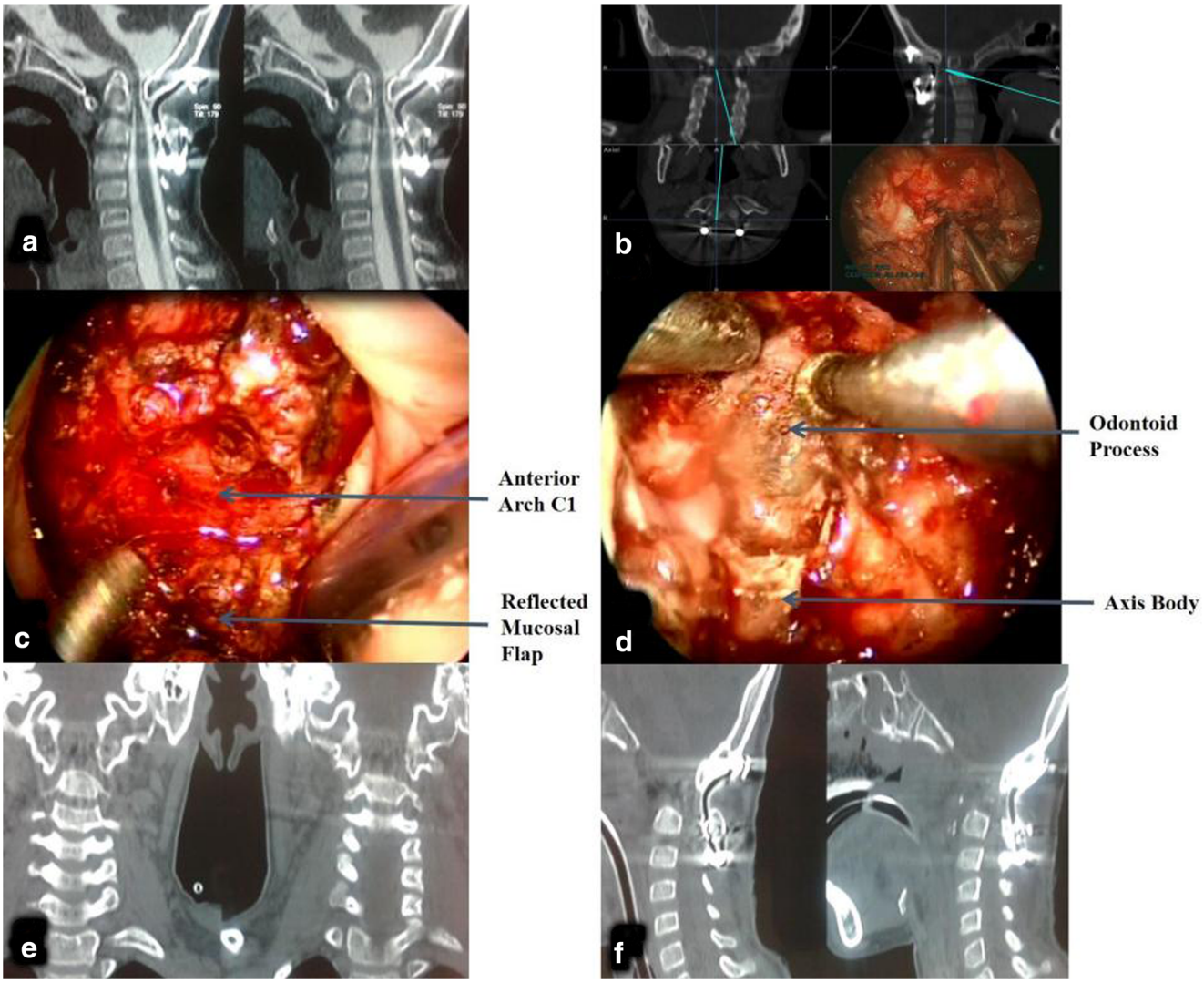

Fig. 7 **a** MRI cranio-vertebral junction of a 4-year old boy with congenital atlantoaxial dislocation and basilar invagination causing cervicomedullary compression. **b** Intraoperative neuronavigation picture taken during transoral odontoidectomy. **c** Intraoperative picture of anterior arch of C1 (blue arrow) and the reflected mucosal flap (blue arrow). **d** Intraoperative picture of the odontoid process (blue arrow) and body of axis (blue arrow). **e** Postoperative CT scan coronal view showing the removed odontoid process. **f** Postoperative CT scan sagittal view showing adequate decompression of the cord with occipito-cervical instrumentation

lower rates of recurrences via transsphenoidal endoscopic route than what has been described in literature previously, although hypothalamic dysfunction and CSF leak still remained significant postoperative morbidities. Locatelli et al. have shown that the endoscopic approach is quite safe for craniopharyngiomas [14] and even for recurrences [15]. Massimi L et al. have shown that endonasal endoscopic surgery for sellar lesions can improve quality of postoperative course in children as compared to sub labial approach [16]. Large cystic craniopharyngiomas presenting with hydrocephalus can be tackled initially with a transcranial endoscopic cyst fenestration followed by definitive endonasal excision of the tumour once the cyst dome collapses after 5 to 7 days [17].

Endonasal endoscopic approach for resection of odontoid process in paediatric population was first described by Kassam et al. [18], and subsequent cases have been described by Beech et al. [19] as well as by Patel et al. [20].

The Pittsburgh group have presented a review of 133 paediatric patients (mean age of 12.7 years) undergoing endoscopic endonasal surgery [21]. They have shown good outcome in almost 90% patient with a CSF leak rate of 10.5%. Stapleton et al. have reported a higher CSF leak rate in a study of 55 children, more so in those with clival chordomas [22].

Shah et al. have shown that pedicled naso-septal flap may be difficult below age of 10 years but is a reliable option over 14 years of age [23]. Purcell et al., in their radiological study, have shown that the nasoseptal flap may not be a limiting factor to repair the sellar defect [24]. Hosan et al. [25] have shown in their study of ten patients that nasoseptal flap may be considered in the first line of treatment for repair. In a review of the

expanded endonasal approach, Khalili et al. [26] have concluded that it is a reliable method of treatment, well tolerated by children.

Conclusion

In spite of the technical challenges posed by narrow passage, small nostrils and ill-developed sinuses in the paediatric age group, surgery from endoscopic transnasal corridor is possible in carefully selected cases of midline skull base lesions with satisfactory achievement of predetermined surgical goals.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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