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Abstract: Background The impact of transnasal endoscopic approaches to the central skull is still increasing. Its use in small children (i.e., under the age of 5 years) is of questionable value. Patients and Methods The present study is a description of a series of five children under the age of 5 years with lesions of the central skull base. Results Three out of five required a combined external and transnasal endoscopic approach. The transnasal access to the central skull base allowed reducing tissue damage and potentially may reduce growth deceleration. Discussion Based on our experience on this small series we suggest that a transnasal endoscopic approach to the anterior and central skull base can be applied even in small children. To successfully operate on such delicate pathologies, a well-working interdisciplinary cooperation is mandatory.

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Is there a place for the endoscope in skull base surgery in children less than 5 years?

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Abstract:

Background:
The impact of transnasal endoscopic approaches to the central skull still is increasing. Its use in small children (i.e. under the age of 5 years) is of questionable value.

Patients and Methods:
The present study is a description of 5 illustrative children under the age of 5 years with lesions of the central skull base.

Results:
3 out of 5. Although 3 of them required a combined external and transnasal endoscopic approach, the transnasal access to the central skull base allowed reducing tissue damage and potentially may reduce growth deceleration.

Discussion:
Based on our experience on this small series we suggest that transnasal endoscopic approach to the anterior and central skull base can be applied even in small children. To successfully operate on such delicate pathologies a well working interdisciplinary cooperation is mandatory.

Keywords:
Endoscopic management; pediatric; skull base surgery; dura plasty; skull base reconstruction.
Background:

Surgery of the central skull base experienced a significant change by the introduction of endoscopic techniques. Several studies could demonstrate that the endoscope allows an excellent illumination and a wide angled view\[1-3]\). These factors seem to compensate the missing three-dimensional view of the microscope as the latter no longer is a significant disadvantage. In addition, the introduction of high density (HD) image quality improved the surgeons view even more so that some spaces in the central skull base can much safer be exposed with the endoscope than with the microscope. Most frequently used rigid telescopes are 4 mm in diameter and thus were considered not to be appropriate for transnasal skull base surgery in children. 2.7 mm telescopes are much more difficult to handle where as the picture quality in times HD image solution and illumination no longer is a major problem.

The majority of studies on transnasal endoscopic skull base surgery in pediatric patients usually comprise children below the age of 18 years without stratification of their age \[4]\). Other surveys are on children and adolescents \[5]\) or children and adults without specific information on the pediatric subset. Larger studies comprised larger series of pediatric patients with lesions of the skalp, skull base and other areas making it impossible to study their specific problems in the endoscopic skull base surgery \[6,7]\.

As a matter of fact, there are only few data published on endoscopic skull base surgery in small children \[8,9]\). It seems to be evident that indications to treat diseases of the skull base through the nostril of small children are rare. We describe a series of children of less than 5 years of age with lesions of the central skull base in which the endoscope provided a significant help to reduce the perioperative morbidity.
Patients and Methods:

There are some publications on frequent diseases like cerebrospinal fluid (CSF) fistula or meningoencephaloceles [8,10,11]. Patients with such pathologies were excluded from this study. In our interdisciplinary skull base surgery unit we have been treating 5 patients of less than 5 years of age over the last 2 years. All underwent preoperative standard contrast enhanced high resolution computed tomography (CT) and magnetic resonance imaging (MR). The individual surgical treatment plan was discussed at the interdisciplinary skull base tumor board. Transnasal tumor resection was performed using a 4 mm rigid telescope with standard HD Camera system (Karl Storz Inc., Tuttlingen, Germany).

Patient 1

This 3 4/12 year old girl was referred with a history of 2 episodes of pneumococcal meningitis within 4 months. Watery discharge or any other nasal symptoms were neither encountered by the parents nor by the treating paediatrician. Preoperative imaging (Fig 1 A and B) showed a widened olfactory fossa on the right and bony dehiscence of the right cribriform plate. Subcranial exposure of the fistula would have implied significant blood loss and the risk for persistent anosmia. On transnasal endoscopic exposure of the right cribriform plate by frontoethmoidectomy as described elsewhere [12] an small arachnoid cyst could be identified (Fig 1C). Removal of the arachnoid cyst was performed using the transnasal transcribriform approach. Sealing of the anterior cranial fossa dura was performed by underlay technique using abdominal rectus fascia and by a pedicled flap from the middle turbinate (Fig 1D). Nasal packing was removed after three days and the girl could be discharged. No meningitis or watery discharge has occurred during 3 years follow-up.
Patient 2

After an uneventful pregnancy of 39 2/7 weeks this boy developed a neonatal respiratory distress syndrome immediately after birth. His birth weight was 3480 g. Due to a bad primary adaptation with Apgar Score 6/4/9 the boy was intubated. Skull sonographic examination showed a large brain tumor requiring further imaging. There, a large in T2 MRI hypointense mass was found compressing almost the entire right hemisphere. Inferiorly, the tumor showed an extension parallel to the pituitary stalk down into the sphenoid and the epipharynx obstructing the nasal airway entirely. This part was partially hyperintense in T2 (Fig 2A). After a biopsy of the epipharyngeal mass the patient developed cerebrospinal fluid (CSF) rhinorrhea. Due to a too small sample size a definitive diagnosis could not be made but the most likely diagnosis was teratoma. Extubation was frustrating as the boy developed respiratory distress with oxygen desaturation. A tracheostomy was performed and the patient was transferred to our institutions. Our first intention was to safe the airway and to close the CSF fistula occurring after the transnasal biopsy. After limited removal of the posterior inferior part of the bony nasal septum (Fig 2B), we removed the tumor part in the epipharynx and sphenoid transnasally (Fig 2C). The stalk like part of the tumor between the intracranial space and the sphenoid could be identified from where CSF was running. The CSF-fistula could be sealed using a pedicled flap from the inferior turbinate. Since then no CSF rhinorrhea was encountered. Pathology remained highly suspicious for teratoma. Three weeks later a pteryonal craniotomy was performed to remove the intracranial part. Intraoperatively the superior part of the inferiorly exposed stalk like part could be exposed and resected. Postoperatively the boy developed a transitory diabetes insipidus and a panhypopituitarism requiring hormone substitution. The latter has been
continued up to present. The final histology revealed surprisingly a glioneuronal heterotopia. Postoperative occurring seizure could be stabilized using topiramat. Postoperative MRI shows neither persistent lesion nor recurrence (Fig 2D). The boy has been decanulated from his tracheostomy after 2 years and he is ambulatory. There is a remaining strabismus, hormone substitution and delayed development.

**Patient 3**

This girl was born after a normal pregnancy period of 39 3/7 weeks. Her birth weight was 3200 g. Primary adaptation was normal. On first clinical investigation a cleft of the soft palate was seen. Moreover through this palatal gap a mass could be identified. Otherwise the girl was healthy and no neurologic deficits could be identified. An MRI in general anaesthesia was performed showing a mass located extradurally between the medial surface of the right temporal lobe and the cavernous sinus. From there the tumor extended down along the sphenoid side wall into the epipharynx as well as into the pterygopalatine fossa (Fig 3 A and B). The girl could not be extubated after the MRI because of significant nasal obstruction and consecutive respiratory distress. A tracheostomy became necessary. On interdisciplinary discussion the removal of the intracranial part of the tumor together with the reconstruction of the upper airway was decided. The transcranial tumor resection was performed through a right orbitozygomatic craniotomy to avoid any retraction of the brain and to gain good access to the cavernous sinus. In the same setting transoral removal of the pharyngeal part through the palatal gap was performed. Histology revealed an immature teratoma. The girl recovered well from this surgical part and grew normally. As the tumor showed a very low proliferation rate on histology and did not significantly grow on follow-up MRI, the remaining tumor mass in the pterygopalatine fossa was planed to remove at the age of
13 months. To remove this part the adjacent paranasal sinuses (i.e. maxillary and posterior ethmoid) were widely opened. After removal of the posterior wall of the maxillary sinus the internal maxillary artery was exposed and ligated. Removing the tumor in piece meal the optic canal superiorly and the cavernous sinus at the level of the foramen rotundum could be identified (Fig 3C). Near total removal of the tumor could thus be achieved with small tumor remnants at the optic nerve. However, there is no tumor left in the more critical area of the cavernous sinus and foramen rotundum (Fig 3D). As we experience a very low growth rate of this immature teratoma we will remove this remnant later using the transnasal approach similar to optic nerve decompression. Most importantly, the patient could be decanulated and does not have any significant neurologic deficit. To remove the tumor in the pterygopalatine fossa from above would have been possible at the same time. However a temporal mobilisation of the zygoma would have been mandatory to access the entire pterygopalatine fossa. Mobilising the zygoma can be associated with growth deficit of the midface.

**Patient 4**

This 4 year old boy has been treated for a small benign tumor of the right orbit and was otherwise healthy. Because of recurrent and intractable headache an MRI was performed showing a mass located in the sella and the upper clivus (Fig 4A). The correlation of the headache with this mass was not certain. A transnasal transclival approach was performed as described elsewhere [13]. Radical tumor removal could be performed. Pathology revealed a mature teratoma. A pertinacious CSF-leak occurring intraoperatively was finally sealed with multilayer technique. Due to the young age we hesitated to use a pedicled mucoperichondrial flap harvested from the nasal septum.
Postoperative imaging confirmed radical removal (Fig 4B). Headache no longer was complained by the patient.

**Patient 5**

At birth of this 3 year old boy a congenital nasal fistula (nasal dermal sinus cyst) was diagnosed and followed clinically. He does not have other diseases and the fistula never got inflamed. As the cystic part started to grow and the boy (Fig 5A) was teased increasingly the parents asked for elimination of the fistula. Imaging showed a fistula that was on the nasal bone in its caudal portion, penetrating the right nasal bone from where it can be followed underneath the nasal bone up to the slightly enlarged foramen coecum (Fig 5A and B). The pit was mobilised and an open rhinoplasty approach was used to mobilise the external part (Fig 5C). The cranial part was removed using an approach like for nasal septoplasty elevating the mucoperichondrium up to the left nasal dome. Here, the fistula could be exposed to its fibrous stalk at the level of the Foramen coecum (Fig 5D). Imaging performed 3 months postoperatively showed no recurrence and a good aesthetic result (Fig 5E). The boy was free of any symptom for one year follow-up.

**Discussion:**

The described series of children below the age of 5 years is very small and their pathologies are very heterogeneous. Endoscopic management of CSF-Leaks and malformations like meningo-encephaloceles from the cribriform plate are already described [14,15]. There are very limited data on tumors involving the anterior skull base in children [16]. Moreover, it was not the objective to compare external approaches with transnasal endoscopic techniques. However, many of similar cases at that age still
will be operated using external approaches exclusively in a more extensive manner. In this particular patient group more extensive external approaches implies more blood loss and more morbidity. Although only few data exist that could prove the lower morbidity of transnasal approaches to the skull base in comparison to the external access Shane and Panizza summarized important facts in their review [17]. Based on namely two investigations [18,19] they conclude that endoscopic transnasal approaches reduce the amount of blood loss significantly. In addition facing the growing skull and the midface in particular every effort has to be considered not to influence the natural growth negatively.

Di Rocco et al. [8] used in his series 2.7 mm and 4 mm telescope without any distinction which one he uses when. In our series we used the 4 mm telescope as it has the advantage of a cleaning system (Clear Vision®, Karl Storz Inc, Tuttlingen Germany) and a grip in contrast to the 2.7 mm telescope. As one of the author's (DH) has experiences of a larger pediatric series, we would state that below 2.2 kg body weight the 4 mm telescope no longer is adequate.

Reconstruction of dural defects still remains the most challenging issue in endoscopic skull base surgery. Castelnuovo and colleagues [20,21] stressed that a careful patient selection is mandatory to successfully reconstruct dural defect endonasally. In accord with Martin et al.[22] they confirm that almost all CSF-Leaks in the anterior skull base area can be sealed endoscopically. According to our experience the more posterior and inferior area of the central skull base management of CSF-leaks are more challenging, in particular in small children. This was particularly of note in the patient (No. 4) with the lesion inferior to the pituitary gland and the upper clivus. It is established from larger
series, that fistula in the sellar region can be difficult to treat[23]. In the posterior and inferior area Shah et al. [24] recommend to use the nasoseptal flap even in pediatric skull base defects. In children this procedure is of questionable value as the child's mucoperichondrium from the nasal septum plays an important role in the growing nose and midface [25]. In addition, the nasoseptal flap is difficult to apply in very anteriorly located dural defect [26]. Although its proven effectiveness the sequelae like permanent anosmia, extensive crusting etc can not be neglected [24,27,28]

Conclusions:

As the transnasal endoscopic techniques proofed to be a minimal disruptive it should be considered in any surgery of the central skull base. Even though external approaches are considered as the approach of choice the transnasal endoscopic technique can be applied in children at any age. Our small series confirm that the 4mm telescope with all its advantages can be used even in newborns. Interdisciplinary preoperative planning is the condition sine qua non of any surgery of the central skull base.

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**Figure legend:**

**Fig 1 A** MRI scan with enlarged olfactory fossa

**Fig 1 B** coronal CT presenting bony dehiscency

**Fig 1 C** endoscopic view of an arachnoid herniation between nasal septum (black asterisk) and middle turbinate (white asterisk)

**Fig 1 D** reconstruction of the dural defect using a pedicled flap from the middle turbinate on to the cribriform plate

**Fig 2 A** coronal MRI scan: with hyperintense stalk like connection between the intracranial and the epipharyngeal portion of the tumor (white arrow heads)

**Fig 2 B** endoscopic exposure of the tumor in the epipharynx (black arrow) after creating a window in the posterior inferior nasal septum (black asterisk) inferior to the middle turbinate (white asterisk)
**Fig 2 C** Endoscopic view after tumor removal. CSF-fistula (black arrow), left pharyngeal ostium of the Eustachian tube (black asterisk).

**Fig 2 D** Sagittal MRI scan left pre- and right postoperatively.

**Fig 3 A** Coronal MRI scan. Tumor lateral to the cavernous sinus (black arrow), orbital apex (white arrow) and in the epipharynx.

**Fig 3 B** Coronal MRI. Tumor in the pterygopalatine fossa close to the optic nerve (white arrow).

**Fig 3 C** Endoscopic view with exposure of the cavernous sinus (black asterisk) and optic canal (black arrow).

**Fig 3 D** Postoperative MRI.

**Fig 4 A** Preoperative MRI scan.

**Fig 4 B** Postoperative MRI scan.

**Fig 5 A** Disfiguring nasal hump by the cystic portion on the nasal bone.

**Fig 5 B** Sagittal MRI left: cystic portion on the nasal bone (white arrow) and solid part underneath the bone lasting to the foramen coecum.
**Fig 5 C** Exposure of the bony nasal dorsum (asterisk) and fistula (arrow) using open rhinoplasty approach.

**Fig 5 D** Endoscopic exposure of the fibrous stalk at the level of the foramen coecum.

**Fig 5 E** Aesthetic result 1 year postoperatively.