# ORIGINAL CONTRIBUTION

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# Spontaneous CSF-leaks and meningoencephaloceles in sphenoid sinus by persisting Sternberg's canal*

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SUMMARY

***Objectives:*** *Cerebrospinal fluid (CSF) leaks and meningoencephaloceles of the lateral recess of sphenoid sinuses are rare findings. A congenital bony defect in the lateral wall of sphenoid sinus called Sternberg's canal could be the origin of these lesions. Their endoscopic transnasal management is challenging though less traumatic than transcranial approaches. The aim of this study was to define Sternberg's canal as a potential source for these rare lesions and to describe their endoscopic endonasal management.*

***Methods:*** *In a retrospective analysis clinical charts of 27 patients with CSF-leaks and / or meningoencephaloceles operated between March 2002 and October 2008 at the University ENT-hospital Graz have been reviewed. All patients were treated by an endoscopic endonasal approach.*

***Results:*** *Five patients (4 female / 1 male) were identified with spontaneous CSF-leaks from sphenoid sinus and meningoencephaloceles. In all five cases, Sternberg's canal was the site of leakage, with the bony and dural defects always located laterally between the maxillary and Vidian nerves. Mean age was 51.2 years and mean body mass index (BMI) was 31.9 kg/m$^2$. All patients were operated using a multilayer closure technique. Two patients had recurrences after 12 days and 7 months, respectively, managed by endoscopic revision surgery resulting in a 100% closure rate after one revision (mean follow-up: 6.5 months).*

***Conclusion:*** *Persisting Sternberg's canal can be the source of spontaneous CSF-leaks and meningoencephaloceles in the lateral recess of sphenoid sinus especially when associated with extensive pneumatisation. Endoscopic management is technically challenging, nevertheless its advantages are a good view of the surgical field while being less traumatic than transcranial approaches.*

*Key words: Sternberg's canal, lateral recess, sphenoid sinus, spontaneous CSF-leaks, endoscopic approach*

## INTRODUCTION

Spontaneous cerebrospinal fluid leaks with meningoencephaloceles restricted to the sphenoid sinus are rare clinical findings [1,2]. As of today, only 17 cases protruding through the so-called Sternberg's canal and extending into the lateral recess of sphenoid sinus, have been described in literature [2-4]. Patients presenting with this special clinical entity usually do not have any history of trauma, tumour or iatrogenic injury. Thus the lesions are considered to originate from a congenital bony defect in the lateral wall of the sphenoid sinus first described by Sternberg in 1888 as the lateral craniopharyngeal canal (Sternberg's canal) [5].

Association between persisting Sternberg's canal, extensively pneumatised sphenoid sinuses, elevated intracranial pressure and obesity are discussed as possible reason for spontaneous rhinoliquorrhea and meningoencephaloceles in this region [6-9].

The aim of this study was to define Sternberg's canal as a potential source for spontaneous CSF-leaks and menignoencephaloceles in this region and to describe their endoscopic endonasal management (Figure 1).

### *Anatomy of Sternberg's canal*

In the embryological period, the future sphenoid bone has cartilaginous precursors namely presphenoid and postsphenoid. The presphenoid includes sphenoid body, lesser wings, and tuberculum sellae. The postsphenoid or basissphenoid forms greater wings, dorsum sellae and pterygoid plates [10].

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Figure 1. Intraoperative snapshot from navigation with the probe pointing at the defect in sphenoid sinus.

Ossification centres appear around the third month of fetal life. Only parts of the medial portion of the pterygoid process are formed by membranous ossification [3].

At the time of birth the ossified presphenoid, lesser wings, basissphenoid, greater wings and pterygoid processes fuse to build the complex sphenoid bone. Only a weak cartilaginous union remains between greater wings, presphenoid and basissphenoid, however corresponding to the future lateral wall of sphenoid sinus. During the neonatal period their bony fusion starts anteriorly [2,3]. If the posterior part fuses incompletely, a bony gap, the so-called lateral craniopharyngeal canal (Sternberg's canal) remains [2,3]. It is located in the posterior part of the lateral sphenoid sinus wall inferior and lateral to the maxillary nerve (V2). After reabsorption of cartilage, Sternberg's canal is closed by connective tissue only, being a point of least resistance at the skull base.

The development of the paranasal sinuses starts at the 3rd and 4th fetal month from invaginations of the mucosa [10,11]. At birth, the sphenoid sinus is not yet visible. In some cases it is only developed as small canal. At the age of 12, it has reached its definite shape, but keeps growing until adulthood [11]. When pneumatisation in large sinuses reaches far laterally a contact between Sternberg's canal and sphenoid sinus can occur [2,3].

Table 1. Grafting material.

| *Patient* | *Material* | *Revision surgery* |
|---|---|---|
| 1 | FL, AF, OX | no |
| 2 | FL, FL, CG, OX | AFBP, FL, AFO, OX |
| 3 | AFBP, FL, CG, OX | AFBP, C, FL, OX |
| 4 | AFBP, FL, FL, OX | no |
| 5 | AFBP, FL, FL, OX | no |

Grafting material used:

AF = Abdominal fat
AFO = Abdominal fat for obliteration
AFBP = Abdominal fat bath plug
FL = Fascia lata
C = Cartilage
CG = Composite graft (mucoperiosteum from middle turbinate)

## PATIENTS AND METHODS

### *Patients*

Between March 2002 and October 2008, 27 (17 m and 10 f) patients suffering from a Cerebrospinal Fluid Leak were operated at the ENT-University Hospital in Graz. Five of these patients (18.5%) showed a spontaneous CSF-leak with meningoencephaloceles in the lateral recess of sphenoid sinus. Four patients were female and one patient was male. The mean age was 51.2 years with a range from 44 to 62 years. All patients were treated via an endoscopic endonasal approach.

In a retrospective study, the following data have been reviewed: Body mass index (BMI), presenting symptoms and duration, previous procedures (if any), defect size, site of defect, surgical approach and technique of defect closure, intraoperative complications, duration of surgery, postoperative complications, length of hospitalization and recurrences (Table 1). Patients were followed endoscopically at regular intervals.

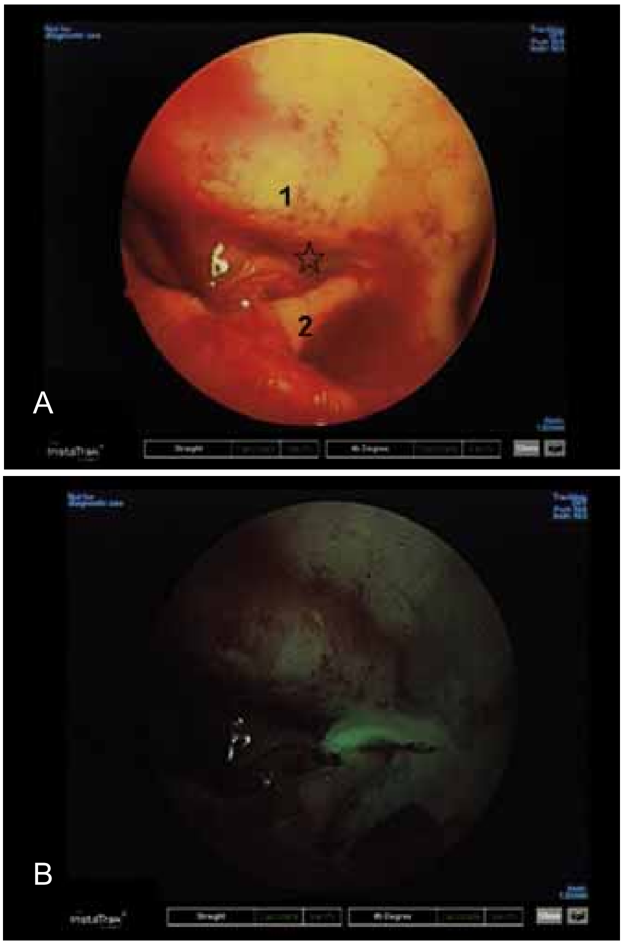


Figure 2. A) Endoscopic view onto defect in right lateral sphenoid recess without bluelight. 1: Bulge of maxillary nerve on its way along lateral sphenoid sinus wall. 2: Vidian nerve in its bony canal. ☆ : Site of defect. B) Endoscopic view onto defect in right lateral sphenoid recess now with bluelight and blocking filter, Na-Fluorescein shining green.

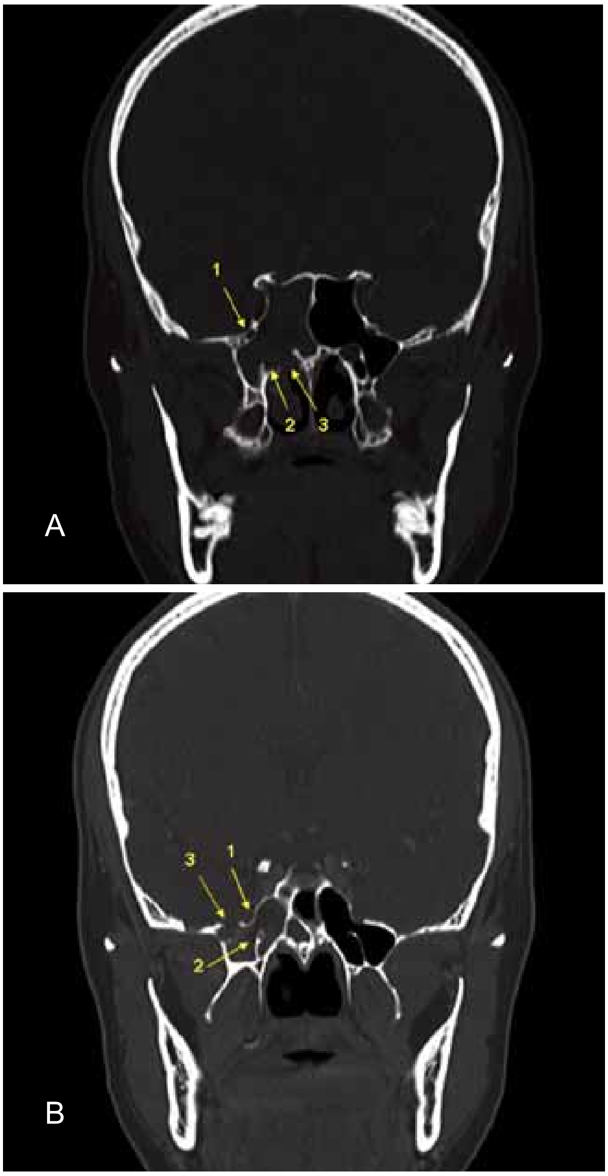


Figure 3. A) Postoperative coronal CT-scan through anterior half of sphenoid sinus. Opacification of sinus from packing material (Oxicell®). 1: Maxillary nerve (V2); 2: Vidian nerve in its bony canal; 3: Resection of sphenoid floor and root of pterygoid process for TESPA. B) Postoperative coronal CT-scan through posterior half of sphenoid sinus. Opacification of sinus from packing material (Oxicell®).
1: Maxillary nerve (V2); 2: Vidian nerve in its bony canal; 3: Sternberg's canal/Site of former leak.

*Surgical technique*

All patients were operated by an endoscopic endonasal approach under image guided navigation (InstaTrak® 3500 Plus, GE Healthcare, Vienna, Austria). On the evening before the operation 0.5 ml to 0.6 ml of a 5% sodium-fluorescein solution were injected by means of a lumbar puncture in all patients. Under blue light, which is attached to the endoscope's light source, sodium-fluorescein shines in a greenish colour whereas all other light is blocked by a blocking filter (Figure 2).

All patients were operated under total intravenous anaesthesia (TIVA) with Propofol and Remifentanyl. No anaesthetic gases were applied. Two surgical approaches were used to reach the defect in the lateral recess of the sphenoid sinus: either a transethmoidal-sphenoidal approach (TESA) or a transethmoidal-sphenoidal-pterygoidal approach (TESPA).

The first steps for both approaches are similar by opening the ethmoid and sphenoid sinuses via a classical sphenoethmoidectomy with preservation of all turbinates. If the defect was located far laterally and below the maxillary nerve (V2), the TESP-approach was applied. Here additionally, maxillary sinus ostium was enlarged and the posterior wall of the sinus was exposed. Then, from the root of the pterygoid process, bone was either resected with a Kerrison punch or drilled away towards the pterygopalatine fossa which was skeletonised. Bleeding from sphenopalatine and maxillary arteries was well avoided or controlled by electrocautery. In addition to a better access to the lateral recess, the maxillary and Vidian nerves were preserved that way (Figure 3).

After having identified the leak, sufficient space was created for access. Mucosa and periosteum around the defect were removed to denude the bone as receptor site for grafts. Meningoencephaloceles were either cauterized or resected. In all five patients a multilayer closure of the defect was performed using fascia lata in an overlay technique (Table 1). In three patients abdominal fat was squeezed into the bony defect in a modified "bath-plug" technique before applying fascia lata (12,13). All layers were additionally fixed with fibrin-glue.
Then, various layers of Oxicell were placed over the defect like shingles filling the entire sphenoid sinus. The surgical field was once again checked for watertight closure under blue light. Finally, some 10 - 20 cm of iodinized gauze were placed into the nose for additional packing.

## RESULTS

At admission to our hospital all patients (n=5) suffered from clear liquid rhinorrhea. Mean duration of consistent or intermittent leakage was 10.6 months (range: 1-30 months). Two patients suffered from periorbital or parietooccipital cephalea, respectively. From documentation available to us no meningitis had occurred.

Patients' mean body mass index (BMI) was 31.9 kg/m$^2$ with a range from 20.55 kg/m$^2$ to 50.39 kg/m$^2$. Three patients were obese with a BMI of ≥ 30 kg/m$^2$ and two patients were within a normal range (20 kg/m$^2$ – 25 kg/m$^2$). None of the patients was previously operated for CSF-leaks. Defect size was assessed in 4/5 cases and measured 5x7 mm, 7x6 mm, 8x10 mm and 4x6mm, respectively (Table 2). All five patients had CSF-leaks with meningoencephaloceles, two located in the left sphenoid sinus and three located in the right sphenoid sinus.

Table 2. Patient data.

| *Patient* | *Gender* | *Age* | *BMI* | *OP-time* | *Defect size* | *Side* | *Approach* | *Postoperative Complications* | *Revision Surgery* |
|---|---|---|---|---|---|---|---|---|---|
| 1 | w | 53 | 34.89 | 2h14 | ? | L | TESPA | no | 0 |
| 2 | m | 62 | 32.88 | 2h44 | 5x7mm | R | TESA | meningitis | 1 (12 days) |
| 3 | w | 44 | 20.55 | 3h05 | 7x6mm | R | TESPA | no | 1 (7 months) |
| 4 | w | 45 | 20.76 | 3h56 | 8x10mm | R | TESPA | no | 0 |
| 5 | w | 52 | 50.39 | 3h42 | 4x6mm | L | TESA | brain abscess | 0 |

In three patients a transethmoidal-sphenoidal-pterygoidal approach (TESPA) was performed. In two patients a transethmoidal-sphenoidal approach (TESA) was chosen.

There were no intraoperative complications. Mean duration of surgery was 188 min (3 hours) with a range from 2h 14min to 3h 56min. No lumbar drainage was applied after surgery. The mean hospitalization was 12 days (range: 10d – 17d).

Endoscopic controls were routinely performed after 1, 3 and 4 months. Mean follow-up time was 6.5 months (range: 1m 3d – 18m 21d). One patient (from abroad) was lost to follow-up, but did not report recurrence within 3 months after surgery.

One patient suffered from temporary numbness on her right upper jaw caused by irritation of the maxillary nerve (V2) which had been exposed during surgery.

Two patients (40%) suffered from recurring rhinoliquorrhea 7 months and 12 days, respectively, after the first surgery. The latter developed bacterial meningitis after revision surgery which was treated successfully with antibiotics.

In both cases revision surgery was performed with a successful closure. During the follow-up period no second recurrence occurred among those two patients. After being uneventful for 4 days, one patient suffered from an epileptic seizure and temporal lobe abscess which were controlled conservatively.

Primary closure rate was 60% in Sternberg's canal leaks (n=5) and 95.5% in all other CSF-leaks (n=25) operated endoscopically at our department during the study period.

## DISCUSSION

"Spontaneous" CSF-leaks occur without any history of trauma, iatrogenic injury, tumour, malformations or radiotherapy [14]. Some of those leaks originate from a bony dehiscence in the lateral sphenoid sinus wall called Sternberg's canal. This special entity of "spontaneous" CSF-leaks is a rare finding. Castelnuovo et al. [4] reported 15 patients with a CSF-leak originating from Sternberg's canal. Blaivie et al. [2] and Schick et al. [3] reported one case each with the same pathology resulting in 17 cases reported in international literature as of today.

Our study includes five patients, thus represents the second largest group in literature. Moreover, those five patients account for 18.5% of all CSF-leaks operated at our department during the study period (Figure 4).

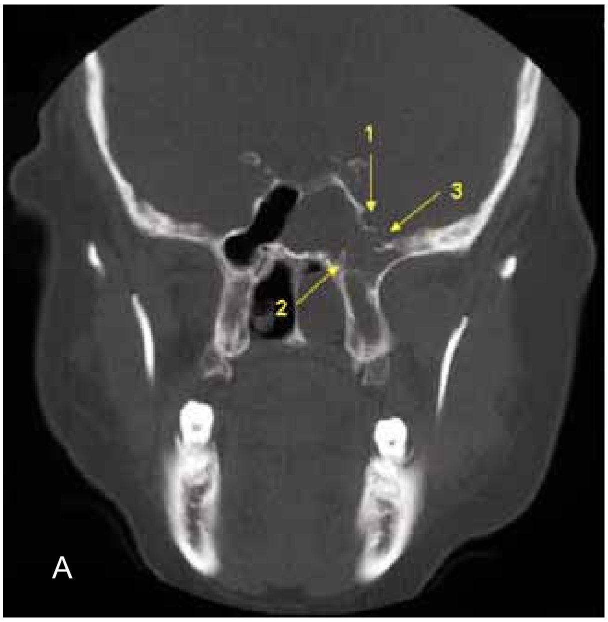



Figure 4. A) Preoperative coronal CT-scan of a left Sternberg's canal CSF-leak. 1: Maxillary nerve (V2); 2: Vidian nerve; 3: Site of leak.
B) Postoperative MRI of same patient with grafting material in situ and mucosal swelling, opacification from packing material in left sphenoid sinus.

A growing number of authors have described endoscopic treatment of CSF-leaks with or without meningoencephaloceles in the sphenoid sinuses [15-18]. Endoscopic treatment increasingly replaces transcranial or extracranial techniques due to its advantages [19]. It is considered to be less traumatic giving a lower morbidity and mortality while providing better access and view of the surgical field. Time needed for surgery, hospitalization and recovery can be reduced and thus costs are decreased [4,15,16,18].

Far laterally extending lesions in the sphenoid sinus can efficiently be reached by the endoscopic transpterygoid approach described by Bolger [20]. In our study, three out of five patients were operated applying that route.

Nevertheless, the endoscopic approach is very challenging, especially when the transethmoidal-sphenoidal-pterygoidal approach is applied. Here one has to open and skeletonise the pterygopalatine fossa encountering the sphenopalatine and maxillary arteries, as well as the maxillary nerve (V2) and Vidian nerve. Careful cauterization of the arteries and preservation of the nerves should therefore always be attempted.

The correct application of intrathecal sodium-fluorescein helps localizing CSF-leaks and checking for watertight closure without any complications [6,7,21,22].

In our study all patients were treated surgically with a multilayer closure of their defects using fascia lata and other materials (Table 1). Some authors use dermal allografts instead of fascia lata [15]. Fascia lata was only applied as an overlay, because of the topographic neighbourhood to neurovascular structures, especially the extremely sensitive temporal lobe. We used abdominal fat in a modified "bath-plug" technique [12,13] to seal the leaks. The plugged-in fat is a primary seal of the leak and supports the adherence of the overlaid fascia lata by reducing the pressure on the fascia exerted from intracranially.

Many authors have looked for explanations for spontaneous CSF-leaks in sphenoid sinuses. Badia et al. [23] and Dunn et al. [24] suggested an association between middle-aged obese women (BMI >30 kg/m$^2$) and spontaneous CSF rhinorrhea. According to Sugerman, central obesity (abdominal trunk) causes higher intraabdominal pressure and thus impairs reflux of blood to the right heart raising intracranial pressure (ICP) [25]. Schlosser et al. state, that an elevated ICP is caused through malabsorbtion of cerebrospinal fluid along the arachnoid villi [6,8,26].

Over the years higher ICP exerts pulsatile forces onto the skull base, especially along areas of least resistance to herniation of dura, further erosion of bone and finally, CSF-leaks [23,24]. In our group four out of five patients were female aged over 40 years and two female patients had a BMI over 30 kg/m$^2$. Mean BMI of the whole group (n = 5) was 31.9 kg/m$^2$. Nevertheless, none of our patients showed indirect signs of elevated ICP.

Another factor associated to spontaneous CSF-leaks in sphenoid sinuses is extensive pneumatisation as described by Shetty et al. [9]. All five patients in our group had extensively pneumatised sphenoid sinuses and prominent lateral recesses in close contact to middle cranial fossa. Sternberg's canal thus represents a site of least resistance in this area [2,3]. Regardless of their cause, spontaneous sphenoid sinus CSF-leaks are difficult to treat.

In two out of five patients (40%) we had recurrences after the first surgical attempt, but a 100% closure rate after one revision surgery so far. One patient had a recurrence at seven months after primary surgery. Compared to these numbers the primary closure rate of all other CSF-leaks operated during the study period is significantly higher with 95.5%. Another study done at our department in 2003 underlines these findings showing a primary closure rate of 84.8%. Here, 33 patients were operated endoscopically for tumours and non-spontaneous CSF-leaks in the paranasal sinuses [27]. Lopatin et al. [19] reported five spontaneous CSF-leaks in lateral sphenoid recess, one of which recurred and had to be revised. All other CSF -leaks (16 in total) in their study did not show recurrence. Gassner et al. [28] reported 95 cases of CSF-leaks treated by different surgical approaches. In their series the endonasal approach was the most successful. Regarding etiology, the highest failure rate (38.5%) after the first surgery was among spontaneous CSF-leaks compared to 26.8% of all CSF-leaks reported in this study. Single site leaks showed the highest failure rate (28.6%) when located in the sphenoid sinus.

## CONCLUSION

A persisting Sternberg's canal should be considered the source of spontaneous CSF-leaks with or without meningoencephaloceles in sphenoid sinuses with extensive lateral pneumatisation, especially when located laterally and below the maxillary nerve.

Endoscopic repair of such leaks is technically very challenging as highlighted by only 60% primary closure rate in our initial series, which compares to a 95% closure rate in all other endoscopic endonasal CSF-leak closures in our department over the last decade. Nevertheless, endoscopic endonasal surgery is safe as no intraoperative complications occurred in our patients. It is less traumatic than transcranial approaches providing a good access and view of the surgical field.

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