

# Expanded Endoscopic Endonasal Approach (EEEA) for Clival Chordomas

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## Abstract

**Objective:** To evaluate the role of expanded endoscopic endonasal approach in removal of clival chordomas. **Patients & Methods:** Nine patients with clival chordomas were operated upon in Cairo University hospital from September 2015 to September 2018 using the EEEA a recurrent case and seven new cases were involved in these study and ten operations were done. All patients had preoperative neurological and radiological examination. The study was focusing on the approach, efficacy of tumor removal, reconstruction of the base and complications related to this approach. **Results:** Nine patients were operated in this study in which ten operations were done. It included six males (66.6%) and three females (33.3%) with age ranging from 4 years to 63 years with average age 40.7 years. Headache and diplopia were the most common symptoms found in six patients (66.6%). Brainstem affection was found in two patients (22.2%). Lower cranial nerves affection was found in two patients (22.2%). One case developed CSF leakage postoperatively (11.1%). Two patients underwent tracheostomy. We achieve total removal in four patients (44.4%), near total removal in one patient (11.1%) and subtotal tumor resection in four patients (44.4%). **Conclusion:** EEEA for clival chordomas is safe and effective approach regarding the results of the incidence of complications, and the percentage of tumor resection.

## Keywords

Endoscopic, Endonasal Approach, Clival Chordomas

## 1. Introduction

Skull base chordomas are locally destructive slowly growing expanding tumors that arise from the remnants of the notochord. They may extend superiorly to the level of cribriform plate of the ethmoids and inferiorly to the craniovertebral

junctions, arch of C1 and to level of 2<sup>nd</sup> cervical vertebrae. It may extend laterally beyond the level of the cavernous sinus and the internal carotid artery and may reach the infra-temporal fossa [1] [2].

Its relation to the brainstem and neurovascular structures makes its removal challenging. Several different approaches have been used as transnasal, transoral, orbitozygomatic subtemporal infratemporal, far-lateral transcondylar and transpetrosal, which have been used for removal of these tumors. Morbidities and mortality may be related to the open approach [3].

With the revolution of technology and improvement of endoscopes regarding their angles, camera and illumination, expanded endoscopic endonasal approach has become one of the most important approaches in removing clival chordomas using the natural corridor of the nasopharynx, decreasing the incidence of morbidity and mortality and expanding the survival rate [4] [5].

## 2. Patients and Methods

A total of nine patients with clival chordomas were operated upon in Cairo University hospitals from September 2015 to September 2018 using expanded endoscopic endonasal approach two patients had undergone previous endonasal surgeries.

**Inclusion criteria:** patients diagnosed with clival chordomas either radiological or histopathological from previous surgery extending superiorly from the cribriform plate and inferiorly up to the level of C2 and fit for surgery.

**Exclusion criteria:** tumor extension behind cavernous sinus, tumor lateral to the internal carotid arteries and tumors with posterolateral extension which require the use of combined approaches and patients unfit for surgery.

### **Preoperative evaluation:**

Computerized tomography (CT) of the paranasal sinuses, sella and clivus was done to detect type of sella, invasion of the bone. Magnetic resonance imaging and angiography (MRI and MRA) were done to detect the relation of the tumor with clivus whether extradural only with affection of the clivus only or of there is intradural invasion and its relation to the brainstem, basal vascular structures and cranial nerves. All cases were operated upon using neuronavigation system and CUSA. All cases received radiotherapy postoperatively.

### **Operative technique:**

All cases were operated upon using the rigid endoscopic by using the four hand techniques by two surgeons (one neurosurgeon) and (one otolaryngologist).

Special instruments are needed in extended endonasal approach as the Mayfield head clamp to fix the head which is important during dissection of the tumor from the brainstem and neurovascular structures.

Different angles of rigid endoscopes are used (0, 30, 70 degrees) to give panoramic view of the field. High special diamond drill to drill the skull base and clivus. Specific long different sizes Kerrison ranger, and different sizes and shapes of suction and dissectors are used due to deep working distance.

The patient is positioned in neutral position with head fixed in Mayfield clamp. Neuronavigation is used in all cases.

**Nasal phase:**

Both nostrils are used to allow introduction of several instruments and the four hand techniques we use. Bilateral middle turbinectomy is done. Complete posterior septectomy is mandatory to widen the field and to prevent dripping of blood from the posterior end of the septum which obscure the field.

Hadad septal flap is prepared pedicled on posterior septal artery for construction later on.

**Sphenoid phase:**

The ostium of the sphenoid sinus is widened with bilateral wide sphenoidotomies up to the medial pterygoid plates laterally.

Sphenoid septum is removal, the floor of the sella is identified and removed with identification of the optic carotid recesses which is the superior limit of our exposure. Bone is drilled in the midline and laterally till the Eustachian tubes which is the lateral limit of our exposure and the landmark for the internal carotid artery. The bone is drilled inferiorly till the lower clivus is reached and craniovertebral junction and the medial part of the occipital condyles which can be removed.

**Tumor removal:**

Extradural tumors involve the bone only and are removed using drill and different types of Kerrison rangers.

In case of intradural tumors, the dura is opened in the midline and tumor is removed by different angles curettes and by KUSA, internal debulking is done and tumor is removed in piecemeal fusion without pulling or using grasping forceps. Safe removal of the tumor is done and if there is tumor adherence to perforators or to the brain stem it should be left without attempt of removal.

**Reconstruction:**

Repair of the dura and reconstruction of the skull base is the most important step in extended endonasal approach to avoid series complications as meningitis which may lead to severe morbidity or mortality.

We reconstruct and repair the dura in all cases even in extradural cases as there are always small defects in the dura during dissection of the tumor.

We use fat for repair of the dura with half of the fat inside the defect and the other half outside the defect to act as a plug and then we fix it with histoacryl.

A second layer of fascia lata is put on lay on top of the fat and sealed with histoacryl to fix it.

Hadad flap is put as a third layer.

Nasal pack is applied for 5-days and removal after that the patient is discharged in the 6<sup>th</sup> post-operative day with instruction of the patent not to lean forward and avoid blowing his nose and constipation for 2-weeks. The average hospital stay is 6 days unless there are complications requiring hospitalization of the patient. Follow MRI was done postoperatively after 3 months, 6 months and then annually.

### 3. Results

Nine patients were operated in our study including six males (66.6%) and three females (33.3%) ranging between the age of 4 years to 63 years with average age 40.7 years. The tumor size ranges from 3 cm to 8 cm with average size (5.5 cm).

**Table 1** shows summary of all patients.

The nine patients underwent ten operations. The average time between the presenting symptom and the time of surgery was 7 months.

Headache and diplopia were the most common presenting symptoms and were found in six patients (66.6%). The headache improved in four patients (44.4%). Diplopia improved in five patients (55.5%). Brainstem affection in the form of hemiparesis was found in two patients (22%). One patient recovered partly with improvement of the weakness (50%) (**Table 2**).

**Table 1.** Summary for patients enrolled.

| Patient no. | Sex    | Age      | Notes          | Symptoms and signs                                   | Intradural extension | Size of tumor | Tumor excision  | Complications   |
|-------------|--------|----------|----------------|--|----------------------|---------------|---|---|
| 1           | Female | 4 years  | Operated twice | Headache and diplopia                                | Present              | >5 cm         | Subtotal Follow-up<br>2 <sup>nd</sup> operation<br>One year → radiation | -   |
| 2           | Male   | 22 years | Recurrent case | Diplopia, Rt side weakness dysphagia                 | Present              | >5 cm         | Subtotal  | iCA injury<br>Temporary tracheostomy  |
| 3           | Male   | 57 years | Recurrent case | Diplopia, Rt side weakness dysphagia                 | Present              | >5 cm         | Subtotal  | CSF leak, meningitis<br>Temporary 7 <sup>th</sup> nerve<br>Permanent tracheostomy and gastrostomy → death |
| 4           | Male   | 32 years | Mo.            | Headache   | No                   | 4.6 cm        | Total removal   | DVT   |
| 5           | Male   | 45 years |                | Headache, diplopia                                   | Present              | 4 cm          | Total removal   | -   |
| 6           | Female | 50 years |                | Hormonal affection                                   | Present              | 4 cm          | Total removal   | -   |
| 7           | Male   | 63 years |                | Headache, diplopia visual affection gait disturbance | Present              | >5 cm         | Subtotal  | Hematoma in fat and fascia lata site  |
| 8           | Female | 55 years |                | Headache   | No                   | 4 cm          | Total   | -   |
| 9           | Male   | 39       |                | Headache, diplopia                                   | Present              | >5 cm         | Near total  | -   |

**Table 2.** Symptoms and signs.

| Symptoms and signs   | No. of patients | Postoperative  |
|----------------------|-----------------|----------------|
| Headache             | 6               | Improvement    |
| Diplopia             | 6               | Improvement    |
| Brainstem affection  | 2               | Improvement    |
| Lower cranial nerves | 2               |                |
| Hormonal disturbance | 1               | No improvement |
| Visual affection     | 1               | Improvement    |
| Gait disturbance     | 1               | No improvement |

Lower cranial nerves in the form of dysphagia were present in two patients (22.2%).

In one patient tracheostomy was done at the same time of surgery as the patient has partial bulbar palsy and the tracheostomy was closed 3-months after operation.

The symptoms of bulbar affection of the other patient were worsened which require the need of permanent tracheostomy and gastrostomy. This patient also had CSF leakage and meningitis which required the insertion of external ventricular drainage and ventriculoperitoneal shunt later on. This patient died from the complication and progression of the disease.

Hormonal disturbance in the form of pituitary insufficiency was found in one patient (11.1%) which didn't improve after surgery.

Visual affection in the form of optic nerve and chiasmal compression was found in one patient (11.1%) which improved after surgery.

Gait disturbance was found in one patient (11.1%) which didn't improve after surgery.

We had two recurrent (22.2%) cases which received radiotherapy after the first operation and were reoperated.

We had one case of a 4-year-old girl which underwent surgery and subtotal excision was done, but the patient didn't receive radiotherapy due to her age and was put under close follow-up and was operated one year later due to progression of the disease followed by radiotherapy.

Only 2 cases were extradural only (22.2%) (**Figure 1**) while the other seven cases had extradural with intradural extension (77.7%) (**Figure 2** and **Figure 3**). **Table 3** shows dural extent.

Five patients (55.5%) had tumor size > 5 cm, while four patients had tumor size < 5 cm with average size (5.3 cm).

We achieve total removal in four patients (44.4%), near total removal in one patient (11.1%) and subtotal removal in four patients (44.4%) (**Table 4**).

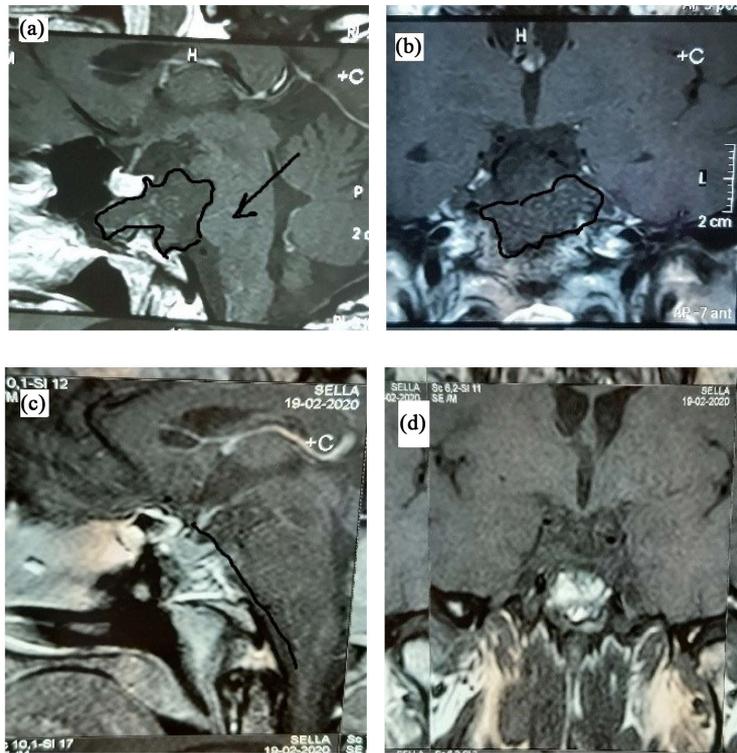
We had one case of intra-operative carotid injury (11.1%) which was controlled by comparison with no subsequent complications, postoperative angiography was done with no pseudoaneurysm detected.

**Table 3.** Dural involvement.

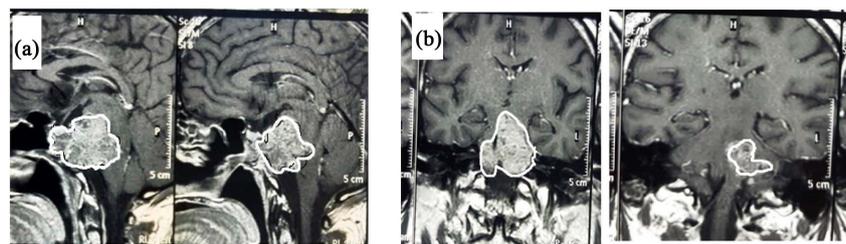
|                      | No. of patients | Postoperative |
|----------------------|-----------------|---------------|
| Intradural extension | 7               | 77.7%         |
| Extradural only      | 2               | 22.2%         |

**Table 4.** Extent of tumor removal.

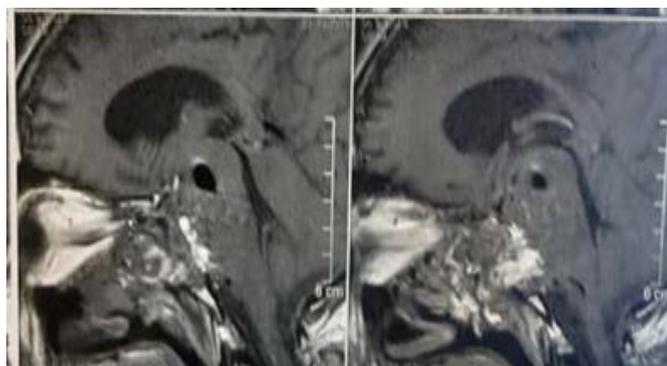
|                          | No. of patients | Percentage |
|--------------------------|-----------------|------------|
| Gross total removal      | 4               | 44.4%      |
| Near total removal > 95% | 1               | 11.1%      |
| Subtotal removal < 95%   | 4               | 44.4%      |



**Figure 1.** MRI of patient number 4 with extradural clival tumor. (a) Preoperative sagittal MRI showing the tumor outlined and the arrow shows the brain stem; (b) Preoperative coronal; Postoperative sagittal (c) and coronal (d) MRI showing total removal of patient number 4.



**Figure 2.** Preoperative MRI sagittal (a) and coronal (b) showing recurrent clival chordoma with intradural extension and invasion of brain stem (patient number 3).



**Figure 3.** Postoperative MRI sagittal showing total removal of the intradural clival chordoma (patient number 3).

We had two cases of tracheostomy postoperatively (22.2%), one was operated at the same time of tumor removal and was closed 3-months later, the other case underwent permanent tracheostomy and gastrostomy due to severe lower cranial nerve affection and the patient died later on.

One case developed cerebrospinal fluid (CSF) leak (11.1%) postoperatively and meningitis and external ventricular drain was inserted followed by insertion of ventriculoperitoneal shunt.

One case developed deep venous thrombosis (11.1%) and was treated by anti-coagulant for 2 months.

One patient developed craniocervical instability (11.1%) due to tumor involvement of arch of C1, part of occipital condyles and odontoid process which required the need to do craniocervical fixation.

Hematoma occurred at the site of fascia lata and fat in one patient (11.1%) which was used for reconstruction.

Hadad flap was done in seven patients (77.7%) but cannot be done in the two recurrent cases (22.2%).

#### 4. Discussion

Clival chordomas are locally destructing bone expanding, slowly growing tumors arising from notochord remnants [6].

The five years survival ranges between 65% and 79% and significantly decreases in 10 years [7].

Several factors determine the approach as the surgeons' experience, familiarity with the approach and extension of the tumor [6].

In the past before the development of endoscopic and its great revolution in the illumination and magnification open surgeries was taking the upper hand which gross total removal (44% - 83%), neurological mobility (0% - 80%), vascular injury (9% - 12%) and CSF leakage (25%) [8] [9].

The first endonasal surgery for clival chordomas was performed by Bouche *et al.* [9]. The first series of EEA to clival chordomas was published by Jhoin (2001) [10].

In our series we operated upon nine patients with the age ranging from 4 years to 63 years with average (40.7 years). It included six males (66.6%) and three females (33.3%).

In the 4 years old girl we operated upon special thin rigid endoscopes and different thin instruments were used due to the small limited field.

In the study done by Martina Stippler [11] which included 20 patients with age ranging from 4 years to 76 years with male to female ratio 2:3.

In our study, headache and diplopia were the most common presenting symptoms and were found in six patients (66.6%). The headache improved in four patients (44.4%), diplopia improved in five patients (55.5%).

The study published by Amir R. Dehdashti *et al.* [12] included 12 patients of which diplopia was found in seven patients (58.3%), improved in five patients (71.4%), and stayed as preoperative in two patients (28.6%).

In the study, published by Giorgio Carrabba *et al.* [13] which included 14 patients, eight patients (57.1%) had diplopia, five patients (62.5%) recovered completely, one partly improved (12.5%) and two patients remain as preoperatively (25%).

We had two cases of brainstem affection in the form of hemiparesis (22.2%), one patient recovered partly with improvement of the weakness (50%). Lower cranial nerves in the form of dysphagia were found in two patients (22.2%). In the first patient temporary tracheostomy was done at the same time of surgery as the patient has partial bulbar palsy and the tracheostomy was closed 3-months after the operation. The symptoms of bulbar affection of the second patient were worsened after the operation which required the need of permanent tracheostomy and gastrostomy. This patient also developed CSF leakage and meningitis which required the insertion of external ventricular drain and ventriculoperitoneal shunt later on. This patient died later on from the sequelae of complication and progression of the disease.

In comparison to the study published by Giorgio Carrabba *et al.* [13] three patients out of 14 patients had lower cranial nerves affection from which two improved and one worsened, one patient with brainstem and long tract affection which improved after surgery.

In the study published by Amir R. Dehdashti *et al.* [12] which includes 12 patients, lower cranial nerves affection was found in three patients, two patients improved and one patient worsened. Brainstem compression symptoms were found in two patients, one patient totally recovered and the other patient remained unchanged.

In our study, we had one case with hormonal disturbance (11.1%) which didn't improve after surgery. Visual affection in the form of optic nerve and chiasmal compression was found in one patient (11.1%) which improved after surgery. Gait disturbance was found in one patient (11.1%) which didn't improve after surgery.

In comparison with the study of Amir R. Dehdashti *et al.* [12] there is one case of optic nerve and chiasmal compression which partly recovered after surgery, one case of anterior pituitary insufficiency which didn't improve after surgery.

In our study, we had two cases extradural only (22.2%), while the other seven cases had extradural with intradural extension (77.7%). Five patients (55.5%) had tumor size > 5 cm, while four patients had tumor size < 5 cm (44.4%) with average size (5.3 cm). We achieved gross total removal in four patients (44.4%), near total removal in one patient (11.1%) and subtotal removal in four patients (44.4%).

In the study, performed by Frank G *et al.* [14] which included eleven patients; total resection was achieved in three patients (33%), subtotal resection in five patients (56%) and partial resection in one patient (11%).

In the study, performed by Martina Stippler [11] which included twenty patients, 12 patients were operated for the first time primary (60%), while the other eight patients were recurrent (48%). Tumor size average size was 29.1 cm<sup>3</sup>.

Intradural extension was found in nine cases (45%).

In the primary cases (12 cases), total resection was achieved in eight patients (66.7%), subtotal resection in two patients (16.7%) and near total resection in two patients (16.7%).

Regarding the recurrent cases, gross total resection was achieved in one case (12.5%), near total resection in two cases (25%) and subtotal resection in five cases (62.5%).

In the study, performed by Giorgio Carrabba *et al.* [13] gross total resection was achieved in (59%) and subtotal resection (>80%) of tumor removal was achieved in (41%).

In the study, performed by Amir R. Dehdashti *et al.* [12] gross total resection was achieved in seven patients (58%), and subtotal resection (>80%) of tumor was achieved in five patients (42%).

In our study, we had one case of intraoperative carotid injury (11.1%) which was in a recurrent care which received radiotherapy. It was controlled by compression with no subsequent complications, postoperative angiography was done with no pseudoaneurysm detected.

One case (11.1%) developed cerebrospinal fluid leakage and meningitis and external ventricular drain was inserted following by ventriculoperitoneal shunt.

One case (11.1%) developed deep venous thrombosis in one case and was treated by anticoagulant for 2 months.

One patient (11.1%) developed craniocervical instability due to tumor invasion of arch of C1 and part of occipital condyles and odontoid process which required the need to do craniocervical fixation.

One case (11.1%) developed hematoma at the site of fascia lata in fat which was taken for reconstruction (11.1%).

We only have one case of mortality due to progression of the disease (11.1%).

In our study, performed by Frank G *et al.* [14] CSF leakage occurred in two patients (22%), vascular injury occurred in one patient (11%), four patients had recurrence (44%) and three patients died from progression of the disease (33%).

In the study, performed by Martina Stippler [11] CSF leakage occurred in five patients (25%) which was decreased after the use of Hadad flap to 5%.

Permanent neurological deficit occurs in one case (5%) due to brainstem hemorrhage, treatment hemiparesis in two cases (10%) and carotid injury in one patient (5%).

In the study, performed by Giorgio Carrabba *et al.* [13] CSF leakage occurred in 24%, tension pneumocephalus in 6% and intracranial hematoma in 6%.

In the study, performed Amir R. Dehdashti *et al.* [12] CSF leakage occurred in four patients (33.3%), in which two resolved by lumbar drain only and two patients required reconstruction and lumbar drain.

One patient (11.1%) developed tension pneumocephalus.

There are certain limitations to our techniques as tumor extension behind cavernous sinus, tumor lateral to the internal carotid arteries and tumors with posterolateral extension which require the use of combined approaches.

All patients received radiotherapy postoperatively, we didn't have proton beam radiation in our country; however, some studies showed that there is no difference in results between the patients that received proton beam therapy in comparison with the patients that received radiotherapy.

The follow-up period which is 14 months is too short and our series of patients is not big, so we recommended a longer follow-up period with a large number of patients.

## 5. Conclusions

EEEA for clival chordomas has become one of the most important approaches to clival chordomas. The safety and efficacy of this approach for removal of midline tumors extending from the cribriform plate superiorly to the odontoid processes inferiorly had made this approach. The approach of choice for these tumors regarding the results, decrease incidence of complications especially vascular complications and CSF leakage, long learning curve, special instrument, and familiarity of the approach is the key point for safe adequate removal of these tumors.

It is appropriate for midline tumors and combined approaches should be used for tumors extending laterally beyond the cavernous sinus and the internal carotid artery.

Our study is limited by the small number of patients and short follow-up period; however, satisfactory outcome had achieved, but we need more numbers of patients and long follow period to detect the exact percentage of recurrence, the complications of the disease itself and the complications related to radiotherapy.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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