# TELOVELAR APPROACH FOR SURGICAL TREATMENT OF FOURTH VENTRICLE TUMORS IN ADULT PATIENTS

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**ABSTRACT**

Purpose: Surgery in and around the fourth ventricle is challenging not only because it has a central location in the Central Nervous System (CNS) but also because it lies in such close proximity to multiple vital structures. Treatment of tumors in this area has nevertheless evolved significantly; with the telovelar, as the most widely used surgical approach today. The aim of this paper is to evaluate results of the telovelar approach for surgical treatment of fourth ventricular tumors, as well as describe the technique. Materials and Methods: A retrospective study was conducted of adult patients undergoing surgery for tumors of the fourth ventricle, between 2011 and 2019 at our institution, using a telovelar approach. Demographic and clinical data were collected together with pre and postoperative test results, operative reports and videos. The Karnofsky Performance Status (KPS) scale was used to measure clinical outcome. Results: During an 8-year period, 18 adult patients with fourth ventricle tumors were operated using the telovelar approach. The most frequent presenting symptom was headache (50%), and the most common pathology reported ependymoma. Gross total resection (GTR) was achieved in 67% of cases and average immediate postoperative KPS score was 86.1, improving slightly during follow-up. Conclusion: Fourth ventricle tumors are relatively uncommon in the adult population. Current treatment continues to be surgical excision and the approach of choice is the telovelar. We present details of the technique together with results of this case series.

**Keywords:** Fourth ventricle; Tumor; Ependymoma; Microsurgery.

**ABORDAGEM TELOVELAR PARA TRATAMENTO CIRÚRGICO DE TUMORES DO QUARTO VENTRÍCULO EM PACIENTES ADULTOS**

Objetivo: A cirurgia dentro e ao redor do quarto ventrículo é desafiadora, não apenas por ter uma localização central no Sistema Nervoso Central (SNC), mas também por estar muito próxima de várias estruturas vitais. O tratamento de tumores nessa área, no entanto, evoluiu significativamente; com o telovelar, como a abordagem cirúrgica mais utilizada atualmente. O objetivo deste artigo é avaliar os resultados da abordagem telovelar para tratamento cirúrgico de tumores do quarto ventrículo, além de descrever a técnica. Materiais e métodos: Foi realizado um estudo retrospectivo de pacientes adultos submetidos a cirurgia para tumores do quarto ventrículo, entre 2011 e 2019 em nossa instituição, utilizando abordagem telovelar. Os dados demográficos e clínicos foram coletados juntamente com os resultados dos testes pré e pós-operatórios, relatórios operatórios e vídeos. A escala Karnofsky Performance Score (KPS) foi usada para medir os resultados clínicos. Conclusão: Os tumores do quarto ventrículo são relativamente incomuns na população adulta. O tratamento atual continua sendo excisão cirúrgica e a abordagem de escolha é a telovelar. Apresentamos detalhes da técnica juntamente com os resultados desta série de casos.

**Palavras-chave:** Quarto ventrículo; Tumor; Ependimoma; Microcirurgia.

## INTRODUCTION

Ventricular tumors are uncommon, corresponding to approximately 1.1% of all central nervous system tumors, and only a small fraction are located in fourth ventricle. [1]

Tumors of the fourth ventricle represent a great challenge for neurosurgeons, because although greater resection is associated with better survival, this in turn needs to be counterbalanced against radicality of tumor removal, with potential morbidity and mortality

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related to inadvertent damage to underlying eloquent structures, including those in the fourth ventricle floor, cerebellar peduncles and deep cerebellar nuclei, as well as risk of vascular damage. [2, 3, 4]

Tumors in this location usually present with signs and symptoms of intracranial hypertension (headache, nausea/vomiting, blurred vision, etc.) or of mass effect, from pressure on the cerebellum and/or brainstem (ataxia, dysmetria, gait instability, diplopia, etc.). [3]

Controversy persists regarding the best approach for fourth ventricle tumor resection. Historically the most common was transvermian, but as a result of high postoperative sequelae including mutism, trunk ataxia, neurocognitive deficit, many neurosurgeons have subsequently opted in favor of the telovelar approach. [2,3,5,6,7,8,9]

The objective of this paper is to present our experience using a telovelar approach for surgery in tumors of the fourth ventricle in adult patients, emphasizing immediate and long-term postoperative results.

## MATERIALS AND METHODS

Retrospective study of 27 adult patients operated for tumors of the IV ventricle at FLENI, Buenos Aires, Argentina, between February 2011 and February 2019. Nine were not operated using telovelar approach, and not included in this analysis.

Demographic data, personal history, clinical presentation, operative report, pathology results and outpatient follow up were obtained from the institutional digital medical record database. Patients diagnosed with swallowing difficulties or lower cranial nerve symptoms preoperatively, underwent laringoscopy and/or videofluoroscopic swallowing exam to objectify degree of neurological involvement.

Brain MRI was obtained in all patients before surgery, and 3 to 6 months after. Images were used to measure maximum tumor diameter, relationship to the floor or ceiling of the fourth ventricle and extension orientation. Degree of resection according to postoperative imaging findings was classified as: Gross Total (GTR), macroscopically complete resection; Near Total (NTR): resection with tumor residue less than 5 mm thick; Subtotal (ST): residue greater than 5 mm. [10]

Parameters monitored during the immediate postoperative period and follow-up included: motor/sensory deficits, cranial nerve function and equilibrium. Number of

hospitalization days was recorded and post-surgical complications evaluated one month after surgery.

Karnofsky Performance Status (KPS) score was used to rate postoperative outcome (Table 1) before discharge and during follow up. [11] Minimum postoperative follow-up was 6 months.

Table 1 - Karnofsky performance Status score. [11]

| Score (category) | Karnofsky |
|---|---|
| 100 | Normal; no complaints; no evidence of disease. |
| 90 | Able to carry on normal activity; minor signs or symptoms. |
| 80 | Normal activity with effort; some signs or symptoms of disease. |
| 70 | Care for self; unable to carry on normal activity or to do active work. |
| 60 | Requires occasional assistance but is able to care for most of his needs. |
| 50 | Requires considerable assistance and frequent medical care. |
| 40 | Disabled; requires special care and assistance. |
| 30 | Severely disabled; hospitalization necessary; active supportive treatment is necessary. |
| 20 | Very sick; hospitalization necessary; active supportive treatment is necessary. |
| 10 | Moribund; fatal processes progressing rapidly. |
| 0 | Dead. |

**Surgical technique for the approach**

All patients were operated in the Concorde position with three-point skull clamp fixation. Correct positioning is essential, particularly degree of cervical flexion, for a safe approach and satisfactory tumor resection.

Motor and somatosensory evoked potentials were monitored in all four limbs, and in facial and lower cranial nerves.

For this technique a median skin incision from 1.5 cm above the inion to C2-C3 is extended at the upper limit to facilitate preparation of a pericranial graft, used for closure in the vast majority of cases. We consider that harvesting this graft during the initial stages of the procedure reduces surgery duration as well as postoperative pain, compared to use of posterior cervical muscle fascia. A medial suboccipital craniectomy tailored to tumor size is then completed. To obtain an adequate working angle with the least possible retraction, the posterior arch of C1 is resected in most cases, particularly when tumors have important caudal

extension, or are located in the upper ¼ of the ventricle. A "Y" or "V" -shaped dural incision, starting from lateral to medial is prepared and the suboccipital sinus sutured or coagulated as needed, trying to preserve the arachnoid surrounding the cisterna magna.

After sampling cerebrospinal fluid from the cisterna magna for cytology, tumor staging and to rule out dissemination along the brainstem, the arachnoid is then opened and the cerebellomedullary cistern dissected along the telo-velo-tonsilo-medullary and uvulo-biventral clefts. In most cases, the tumor can be seen protruding at this stage, if not, the tela choroidea and the inferior medullary velum are opened. This procedure is unilateral, and when necessary, extended upwards or to the contralateral side. Once the tumor is identified, a biopsy is taken for intraoperative study and the margins dissected until the floor of the fourth ventricle is reached. The latter is protected with cottonoids, as is the upper pole, to prevent obstruction of the aqueduct with blood clots and detritus. Microsurgical piecemeal tumor resection is performed using an ultrasonic aspirator. On reaching the implant site, in cases where a clear plane of cleavage between the tumor and the ventricle floor is not identified, a thin layer of tumor is left in situ, in an attempt to improve functional outcome.

## RESULTS

### Patient and tumor characteristics

Table 2 shows the characteristics of the 18 patients, who underwent surgery, average age was 37.5 years (range 21-72 years); male: female ratio 2.8 and average postoperative follow-up duration 36.5 months (range 6 - 85 months).

Table 2 - Patient characteristics

| TABLA 1 | | |
|---|---|---|
| Characteristic | | Nº of patients |
| Sex | | |
| | Female | 5 |
| | Male | 14 |
| Presenting symptom | | |
| | Headache | 9 |
| | Gait instability | 7 |
| | Nausea/vomiting | 6 |
| | Limb ataxia | 3 |
| | Cranial nerve deficit | 3 |
| | Vertigo | 1 |
| | Neuralgia | 1 |
| | Disorientation | 1 |
| | Blurry visiom | 1 |
| | Dysphagia | 1 |
| | Asymptomatic | 2 |
| Implant site | | |
| | Floor | 13 |
| | Ceiling | 3 |
| | Cerebellar peduncles | 2 |
| Extension | | |
| | Caudal | 13 |
| | Posterior | 5 |
| | Lateral | 3 |
| | Cranial | 2 |
| | Anterior | 1 |
| | Limited to IV ventricle | 1 |
| Pathology | | |
| | Ependymoma | 6 |
| | Subependymoma | 3 |
| | Choroid plexus papilloma | 3 |
| | Rosette-forming glioma | 2 |
| | Medulloblastoma | 2 |
| | Hemangioblastoma | 1 |
| | Epidermoid | 1 |

Preoperative imaging analysis showed average tumor diameter was 32 mm (range 16-50 mm), average tumor location was 13mm from the floor of the fourth ventricle, 3 from the ceiling and 2 from the cerebellar peduncles. The majority presented floor implantation and caudal extension. None presented lesions at other CNS sites.

The most frequent symptom was headache (50%), followed by gait instability (39%) and nausea/vomiting (33%). Only 3 patients (16.6%) presented cranial nerve deficits, and 6 patients presented hydrocephalus at diagnosis, with multiple symptoms. Average pre-surgical KPS score was 87.2 points (range 40-100).

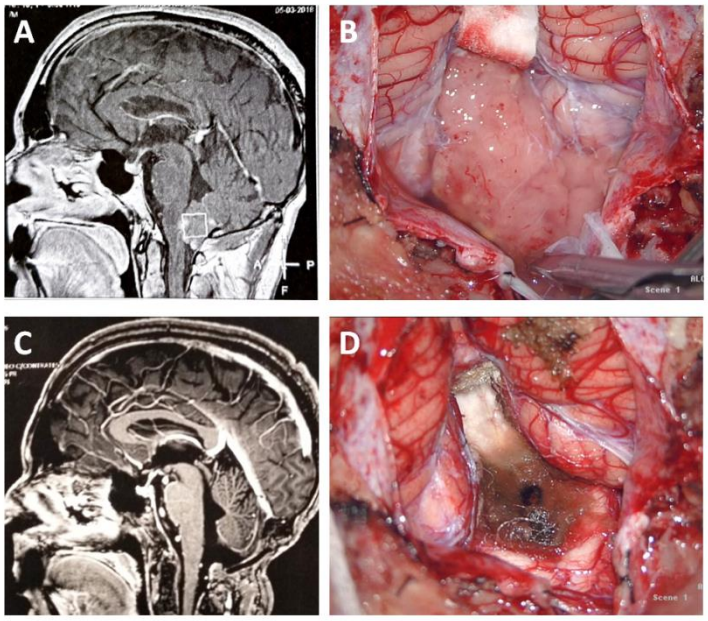
The most common tumor was ependymoma (27.7%), followed by subependymoma and choroid plexus papilloma (16.6% both).

**Illustrative Case**

A 50 year-old male patient consulted for dysphagia. On physical examination no objective neurological deficits were detected. MRI showed a non-contrast enhancing-tumor originating from the inferior medullary velum, with areas of calcification. Surgical resection was performed using a telovelar approach. Pathology report confirmed diagnosis of

subependymoma. The patient evolved favorably, and currently has a KPS score of 100 points. (Image 1)

Image 1 - Subependymoma of the inferior medullary velum: A) Pre-surgical MRI, T1 sequence with gadolinium; B) Intraoperative imaging prior to resection; C) Post-surgical MRI, T1 sequence with contrast; D) Image of the surgical bed after excision and placement of hemostatic material




**Extent of resection and procedure duration**

GTR was obtained in 67% of cases, STR in 22 % and NTR in 11%. Average duration of surgery was 4.4 hours (range 3.25 - 7 hrs).

**Follow-up**

Average hospital stay was 10.5 days (4 - 28 days), only 2 patients (11%) presented postoperative cranial nerve deficit and 3 (16.6%) presented trunk ataxia at discharge. Complications observed included: 3 patients with cerebrospinal fluid (CSF) leak (16.6%), 2 pseudomeningoceles (11%) and 3 (16.6%) cases of meningitis. Six patients required a second surgery, 3 for CSF leak repair and wound closure (16.6%) and 3 (16.6%) for ventriculoperitoneal (VP) shunt placement. KPS score was 84.4 points on average (range 40-90) at discharge, and 92.2 at follow-up (range 40-100).

Only one (6.5%) patient underwent re-resection for tumor recurrence, 2 (16.6%) received chemotherapy and 6 (33%) radiation therapy.

## DISCUSSION

Historically, tumors of the fourth ventricle were operated using the transvermian approach, but as a result of associated morbidities, mainly balance disorders and mutism [5, 12], its popularity began to wane. The telovelar approach was initially described by Matsushima et al. in 1992 [13] and has been extensively studied by other authors since then. [14,15] In the literature, most studies on this subject address treatment of a particular CNS pathology, such as ependymomas or medulloblastomas, and not overall results of fourth ventricle tumor resections, complications, or type of approach used.

The transvermian approach causes damage to functional CNS tissue, and may require resecting the tonsils or part of the vermis; while the telovelar, takes advantage of an anatomical corridor. Tanriover et al. compared both approaches in relation to the degree of exposure of the fourth ventricle offered by each technique, concluding that the transvermian provides a better working angle in the sagittal plane at the level of the superior ¼ of the fourth ventricle, while the telovelar offers additional exposure of the superolateral recess and the Luschka foramen. [15] In this series, most tumors had an inferior extension, which made the telovelar approach an ideal choice.

Currently, the telovelar approach is used extensively around the world, [3,5] and several large series on the treatment of fourth ventricle tumors have been published including ones by Han et al. [16], Tomasello et al. [5], and Ferguson et al. [3] In this study from a center in Latin America, degree of postoperative deficit recovery was expected to be lower since only adult patients were analyzed, which in turn led local surgeons to be somewhat more conservative in terms of fourth ventricle floor resection boundaries.

Morbidity nevertheless remains high, mainly at the expense of gait and swallowing disorders, as well as cranial nerve deficits secondary brainstem involvement. [2, 3, 17], and one study on cerebellar mutism after posterior fossa tumor resection in children, did not find the telovelar approach to be superior in relation to this complication. [18] For these reasons, the jury is still out on which is the best approach for surgery in or around the fourth ventricle, ultimately depending on surgeon preference.

In our practice, we generally prefer the telovelar approach because postoperative morbidity is usually less and normal structures are better protected, while still providing good access to the tumor and early exposure of the floor of the fourth ventricle. As mentioned above, when no clear surgical plane of cleavage between the tumor and the floor of the fourth ventricle is present, a thin layer of tumor remnant is left in situ. In this series 13 of the 18

patients had tumors implanted on the floor of the fourth ventricle, explaining in part why GTR was not achieved in 33% of cases. Our goal was to offer patients acceptable overall survival with good long term quality of life, striking a balance between local disease control and postoperative neurological deficits.

The prevalence of headache as chief complaint reflects the fact that most symptoms are secondary to intracranial hypertension. Only 3 patients of the 6 with hydrocephalus at time of consultation required a VP shunt, suggesting it may be preferable not to rush the decision of placing the device.

The most frequent tumor diagnosed was ependymoma, as in other published series [3], in contrast with pediatric cases, in which medulloblastomas are more predominant. [18, 19]

During postoperative follow-up, despite performing watertight closure of dural defects using autologous grafting, 16.6% of patients developed postoperative CSF leak, a figure slightly higher than that of other series. [3,8] In relation to postoperative need of VP shunting, the rate was 16.6%, lower than in pediatric series where it can reach 30% [8] but higher than in other adult patient series. [3,5] Finally, the average follow-up was relatively short to draw conclusions on disease control, but only 1 patient required a second surgery, 46 months after the initial subtotal resection because of tumor adhesion to the fourth ventricle floor.

Limitations to this study include: 1) selection bias arising from the fact that at our institution, approaches such as the transvermian are often reserved for re-operations in patients with disease recurrence and therefore with potentially worse results in terms of postoperative morbidity; 2) the series included adult patients exclusively, limiting interpretation of findings to this age group; 3) this was a retrospective study and the data was limited to information in medical case files; and 4) although focused on results related to the approach, it is clear that tumor pathology is more relevant regarding long-term results.

## CONCLUSION

Fourth ventricular tumors are relatively uncommon in adults. At present, the treatment of choice continues to be surgical resection. Although treatment-related morbidity remains high and controversy persists regarding the ideal approach, the telovelar is ideal, especially in patients without prior surgery. We present results of a case series, and describe the technical points we consider of importance to improve outcomes.

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