



Original Investigation

Telovelar Approach to Pediatric Fourth Ventricle Tumors: Feasibility and Outcome

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ABSTRACT

AIM: To describe our institution's experience with the telovelar approach as well as factors governing radicality and outcome for pediatric fourth ventricle tumors.

MATERIAL and METHODS: During March 2015-January 2017, 44 children with fourth ventricular tumors were operated using the telovelar approach. Radicality, neurological outcome, complications, and survival rates were evaluated. Statistical analysis was performed to determine factors associated with radicality and neurological outcome.

RESULTS: Gross tumor removal was achieved in 37 patients (84.1%). All patients required cerebrospinal fluid (CSF) diversion through a ventriculoperitoneal shunt. Six patients (13.6%) developed cerebellar mutism, 4 patients (9.1%) developed bulbar paralysis (lower cranial nerve palsy), and 13 patients (30.2%) had tumor recurrence. Medulloblastoma was associated with the highest risk of cerebellar mutism (p=0.040). A poor prognosis (according to overall and progression-free survival rates) was associated with subtotal resection (p=0.020), large cell/anaplastic medulloblastoma and anaplastic ependymoma (p=0.038), highrisk medulloblastoma (p=0.005), and CSF seeding (p<0.001).

CONCLUSION: The telovelar approach provides an adequate anatomical exposure of the fourth ventricle and allows early visualization and protection of its floor (brainstem). It facilitates adequate radicality and is associated with a lower incidence of cerebellar mutism and other approach-related complications. The working angle, particularly to the rostral part of the fourth ventricle, can be increased by removing the posterior arch of the atlas.

KEYWORDS: Pediatric, Fourth ventricle, Posterior fossa, Telovelar approach, Cerebellar mutism

■ INTRODUCTION

ts direct relation to the brainstem and deep location anterior and ventral of the cerebellum make access to the fourth ventricle challenging (24). The transvermian approach to the fourth ventricle with splitting of the inferior vermis may result in caudal vermis syndrome, cerebellar mutism, or injury of the dentate nuclei resulting in head nodding, truncal ataxia, and nystagmus (2,11,21,30).

The telovelar approach can be used as the standard approach for almost all fourth ventricle lesions. However, to date, the clinical series reporting on the pros and cons of this approach do not provide sufficient data to adequately evaluate the outcome (26).

MATERIAL and METHODS

This prospective study included 44 male and female patients with newly diagnosed fourth ventricular tumor treated at

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Cairo University Children's Hospital, Egypt, during March 2015-January 2017. Patients with exclusively cerebellar or cerebellopontine angle lesions, recurrent, and residual lesions were excluded. All legal quardians of patients provided written informed consent.

The patients were assessed for their ventricular size (hydrocephalus) through preoperative imaging [computed tomography (CT) and magnetic resonance imaging (MRI)]. All patients had obstructive hydrocephalus, for which ventriculoperitoneal (VP) shunts were inserted.

Routine removal of the posterior arch of the atlas approximately 0.5 cm from the midline on each side was performed in all cases before the occipital bone flap was elevated. The steps of the telovelar approach were followed with dissection of the arachnoid membrane bilaterally along the tonsillomedullary and uvulotonsillar fissures, allowing for segregation and safer retraction of the cerebellar tonsils (unlocking of tonsils). This gave access to the tela choroidea, which was opened by sharp dissection before the inferior medullary velum was incised. Performing these steps bilaterally allows wide access to the entire fourth ventricle and total tumor resection. In large tumors, central debulking was performed first. The tonsils were retracted slightly upward using retractors. Early identification of the tumor-brainstem interface enabled the protection of the brainstem with a cottonoid. Total tumor excision was the primary goal of surgery, leaving a thin tumor sheet on the ventricle floor only in cases with brainstem invasion (without entering the brainstem). Head flexion together with atlas arch removal facilitated the exposure of the entire length of the ventricular floor up to the Sylvian aqueduct.

Postoperatively, all patients underwent a full neurological examination with an emphasis on posterior fossa syndromes. particularly whether they had complete or partial cerebellar mutism syndrome with determination of the exact components involved (linguistic, neuro-behavioral, and/or motor). Additionally, the examination assessed any delayed cerebrospinal fluid (CSF) collection and CSF leaks.

Non-contrast CT of the brain was performed for all patients within 24 hours of the operation to exclude hematoma and significant pneumocephalus and to assess ventricular size. MRI of the brain was performed within 48 hours to evaluate the extent of tumor resection. During follow-up, MRIs of the brain and spine without and with contrast were performed every 3 months in the first year and every 6 months thereafter to detect any tumor recurrence and/or CSF seeding.

Histopathological examination of the biopsy material and grading according to WHO classification were performed for all patients. Cytological examination was performed on CSF samples obtained no earlier than 2 weeks postoperatively to assess CSF seeding.

All patients, except those with pilocytic astrocytoma grade I (GI), received adjuvant radio- and/or chemotherapy according to their histopathological diagnosis and their risk stratification in case of medulloblastoma.

The outcome was assessed in all patients on the following occasions: immediately postoperative, at discharge, 3 weeks postoperatively, and every 3 months thereafter.

SPSS program (SPSS Inc., Chicago, IL, USA) was used for statistical analysis. Overall survival (OS) was defined as the duration (in months) between tumor removal and final followup or death. Progression-free survival (PFS) was defined as the duration (in months) between tumor removal and first relapse at any site, final follow-up, or death.

The following factors were evaluated with regard to their prognostic value: age (<5 vs. 5-18 years), gender, metastatic stage (M0 vs. M+), extent of resection [gross tumor resection] (GTR) vs. subtotal tumor resection (STR)], risk stratification of medulloblastoma [(MB) (standard risk (SR) vs. high risk (HR)], pathological type, tumor site (with vs. without fourth ventricle floor infiltration), and tumor size (<5 vs. >5 cm in maximal diameter). Survival analysis (PFS and OS) was performed for the various outcome factors (univariate analysis) using Kaplan-Meier statistics. Cox multivariate regression was used to determine the effect of important variables on OS and PFS. p<0.05 was considered statistically significant.

■ RESULTS

Patients Characteristics

There were 27 male and 17 female patients (1.6:1) (Table I); the chief presenting symptoms and signs are listed in Table II.

Patients aged 5-18 years showed no significant difference in PFS from those aged <5 years (2-year PFS 70.6% vs. 62.7%, p=0.508) (Table III).

Tumor Characteristics

Tumors were classified according to their size (maximal diameter in cm) into two groups: <5 cm and >5 cm as shown in Table I.

Tumor Location and Extent of Resection

Tumor location was defined according to preoperative imaging and surgical reports. The tumors were divided into two subtypes (Table I). The first subtype comprised tumors with fourth ventricle (V4) floor infiltration (attachment to brain stem); the second subtype comprised tumors without V4 floor infiltration. The two groups showed no statistically significant difference in outcome (p=0.947 for PFS and p=0.690 for OS). GTR was defined as either a tumor residue <1.5 cm² in the postoperative MRI, no visible tumor remaining or a thin layer of tumor left on the brain stem. Of the 24 patients whose tumors were found to invade or be attached to the brainstem, GTR could be achieved in 17 patients (70.8%). In the remaining patients (29.2%), STR was performed because complete resection was prevented by tumor attachment to or invasion of the brainstem (p=0.008) (Table I). Patients who underwent GTR had a better outcome than those who underwent STR (2-year PFS 93.3% vs. 18.7%, p=0.020).

Histopathological Types

The pathological subtypes of the tumors in our study are listed in Table I. Patients with pilocytic astrocytoma (AS) Grade II (GII), and ependymoma (EP) GII had no progression during the follow-up period and the best outcome (2-year PFS 100.0%). Patients with classical medulloblastoma (CMB) exhibited a better PFS and OS than those with desmoplastic-nodular medulloblastoma (DNMB; 2-year PFS 66.0% vs. 60.0% respectively, p=0.038). Large cell/anaplastic medulloblastoma (LC/A MB) and EP Grade III (GIII) patients had the worst prognosis (2-year PFS 33.3%).

Table I: Clinical, Demographic and Surgical Data of Study Cases

Characteristic	No. of patients	
	n (%)	
Age (years): Mean; 6.05 (range 2 to 15)		
<5 years	17 (38.6)	
5-18 years	27 (61.4)	
Gender	27 M / 17 F	
Tumor Size		
<5 cm	15 (34.1)	
>5cm	29 (65.9)	
Tumor location		
With V4 floor infiltration	24 (54.5)	
Without V4 floor infiltration	20 (45.5)	
Extent of tumor resection		
GTR	37 (84.1)	
STR	7 (15.9)	
Pathological subtypes		
Classic (CMB)	14 (31.8)	
Desmoplastic/nodular (DNMB)	5 (11.4)	
Large cell/anaplastic (LC/A MB)	8 (18.2)	
Ependymoma GII	4 (9.1)	
Anaplastic ependymoma GIII	3 (6.8)	
Pilocytic GI (PA GI)	9 (20.5)	
Grade II, with piloid features	1 (2.3)	
Risk stratification of medulloblastoma		
Standard (Average) risk (SR)	14 / 27 (51.9)	
High risk (HR)	13 / 27 (48.1)	

F: Female, GTR: gross total resection, M: Male, STR: subtotal resection, V4: fourth ventricle.

Postoperative Complications

The main postoperative complications were (in decreasing order of frequency) neck pain, pseudomeningocele, CSF infection, cerebellar mutism, bulbar palsy, and subdural collection (Table II).

CSF infection occurred in seven patients (15.9%). They were managed by shunt removal and external ventricular drain insertion until infection subsided, followed by insertion of a new VP shunt. One patient developed severe ventriculitis and septicemia and died.

Bilateral subdural fluid collection occurred in four patients (9.1%): two were managed conservatively and two required evacuation and upgrading of their medium pressure VP shunts to high pressure shunts.

Cerebellar Mutism Analysis

Cerebellar mutism (CM) occurred in six patients (13.6%), and all of them had medulloblastomas (four patients had CMB and two had LC/A MB). The onset of mutism varied from 2 to 3 days postoperatively [mean: 2.50 ± (standard deviation) 0.548 days].

Table II: Clinical Data of Study Cases

Charactristic	No. of patients (%)
Presenting Symptoms and	Signs
Hydrocephalus	44 (100.0)
Headache & Crying	43 (97.7)
Abnormal gait	31 (70.5)
Vomiting	30 (68.2)
Papilledema	29 (65.9)
Blurring of Vision	28 (63.6)
Diplopia	14 (31.8)
Cranial nerve palsy	8 (18.2)
Head tilt	8 (18.2)
Bulbar symptom	3 (6.8)
Ataxia	2 (56.8)
Mainly encountered postor	perative complications
Neck pain	19 (43.2)
Psedomeningocele	12 (27.3)
CSF infection	7 (15.9)
Cerebellar Mutism	6 (13.6)
Bulbar palsy	4 (9.1)
Subdural collection	4 (9.1)

Four patients (9.1%) had mild to moderate mutism with a decrease in verbal output, hypotonia, and oropharyngeal dysfunction, leading to dysphagia and decreased oral intake. In these four cases, gradual spontaneous improvement occurred within 2-6 months postoperatively, with residual dysarthria. The other two patients (4.5% of all patients) suffered from severe posterior fossa syndrome with all three components (linguistic, motoric, and neurobehavioral). These two patients had LCA MB and died from disease progression later (6 and 8 months after excision, respectively).

Univariate analysis showed a statistically significantly higher risk of CM for medulloblastomas than for other tumor types (p=0.040), but no significant differences were observed for age (p=0.380), gender (p=0.662), tumor size (p=0.394), and the extent of resection (p=0.586).

Postoperative Bulbar Palsy Analysis

Three patients (6.8%) had bulbar symptoms starting preoperatively and four (9.1%) had new-onset bulbar symptoms. Of them, five had LC/A MB and two had EP GIII. Two patients had mild to moderate choking that improved

Table III: Two-Year PFS and OS Univariate Analysis in the Different Subgroups

Characteristics	PFS (%) (Time/month)	р	OS (%) (Time/month)	р
Age (n=43)				
<5y (n=17)	62.7 ± 1.2% (14.686)	0.508	73.2 ± 1.2% (16.353)	0.883
5-18y (n=26)	71.8 ± 9.1% (16.273)		74.7 ± 9.0% (16.987)	
Gender (n=43)				
Male (n=26)	66.3 ± 9.9% (15.595)	0.001	77.1 ± 9.1% (17.309)	0.428
Female (n=17)	70.6 ± 11.1% (15.647)	0.961	70.6 ± 11.1% (15.882)	
umor location (n=43)				
Subtype 1 (n=23)	69.0 ± 9.8% (15.638)	0.947	77.1 ± 9.1% (17.026)	0.690
Subtype2 (n=20)	67.2 ± 11.1% (15.627)		70.1 ± 1.1% (16.336)	
Extent of Resection (n=4	13)			
GTR (n=37)	73.8 ± 7.6% (16.529)		78.3 ± 7.4% (17.933)	0.065
STR (n=6)	33.3 ± 19.2% (7.500)	0.020*	50.0 ± 20.4% (11.833)	
Metastatic stage (n=43)				
M0 (n=28)	93.3 ± 6.4% (18.857)	<0.001*	96.4 ± 3.5% (19.393)	<0.001*
M+ (n=15)	18.7 ± 11.4% (8.347)		31.4 ± 13.7% (11.059)	
MB Risk				
HR (n=13)	21.5 ± 13.0% (8.708)	0.005*	26.2 ± 14.7% (9.890)	0.000*
SR (n=14)	85.1 ± 9.7% (17.845)	0.005*	92.3 ± 7.4% (19.000)	0.003*
Pathology (n=43)				
CMB (n=14)	65.7 ± 14.4%		71.8 ± 1.4%	
DNMB (n=5)	60.0 ± 21.9%		80.0 ± 17.9%	
EP GIII (n=3)	33.3 ± 27.2%		66.7 ± 27.2%	
LC/A MB (n=8)	33.3 ± 18.0%	0.038*	33.3 ± 1.80%	0.044*
EP GII (n=3)	100.0%		100.0%	
PA GI (n=9)	100.0%		100.0%	
AS GII (n=1)	100.0%		100.0%	

AS GII: Astrocytoma grade II, CMB: Classic Medulloblastoma, DNMB: Desmoplastic nodular medulloblastoma, EP GII: Ependymoma grade II, EP GIII: Ependymoma grade III, GTR: Gross Total Resection, LC/A MB: Large Cell Anaplastic Medulloblastoma, MO: No CSF seeding, M+: Positive CSF seeding, OS: Overall Survival, PFS: Progression-Free Survival, STR: Subtotal Resection, V4: Fourth Ventricle, *= Statistically significant.

gradually over 1 month. The other five patients required tracheostomy, which was discontinued 2–4 months later. The univariate analysis showed a significantly higher risk of bulbar paralysis in patients who had undergone STR than in those undergone GTR (p=0.034). Tumor size and histopathological type had no significant impact on developing bulbar palsy (p=0.737 and p=0.116 respectively).

Outcomes

Risk stratification of medulloblastoma

Medulloblastoma patients were divided into two groups: High-risk (HR) defined as M+ Chang stage (metastatic), age <3 years old and/or residual tumor >1.5 cm²; and average/ standard-risk (SR): no metastasis or residual tumor. The SR patients had more favorable PFS than the HR patients (2-year PFS 85% vs. 21.5%, p=0.005). Nine of 13 HR patients (69.2%) experienced a relapse during the follow-up period, whereas two of 14 SR patients (14.3%) relapsed (Table I).

Disease progression and mortality

Tumor metastasis (tumor seeding): the patients were classified into M0 (without CSF metastasis) or M+ (with CSF metastasis) Chang stages according to their CSF cytology and/or MRI of the cranio-spinal axis. The M0 group had statistically significantly better PFS and OS than the M+ group (2-year PFS 93.3% vs. 18.7%, p<0.001). In total, 29 of 44 (65.9%) patients had no metastasis (M0). Fifteen patients (34.1%) had metastases, two of these patients had spinal seeding (enhanced nodules) before excision, 10 had positive CSF cytology, and three developed leptomeningeal dissemination later on. Four of the 14 CMB patients (28.6%), two of the five DNMB patients (40.0%), all LC/A MB patients, and one of the three patients (33.3%) with EP GIII had metastases. Time to metastasis was 0–15 (mean: 0.80 \pm 1.821) months.

Tumor recurrence: Three patients (6.8%) had a tumor recurrence, two with EP GIII (recurrence 2 and 6 months after excision, respectively, the patient with early recurrence died 1 month later), and one patient with CMB (recurrence after 5 months). The time to recurrence was 2–6 (mean: 4.33 ± 2.082) months.

Disease progression: Patients with pilocytic astrocytoma, AS GII, and EP GII had no progression during the follow-up period and overall the best outcome. Thirteen patients (30.2%) had disease progression: two patients had tumor recurrence, ten had tumor metastasis, and one had both. These thirteen patients exhibited the following tumor types: four of fourteen CMB patients (28.6%), two of five DNMB patients (40.0%), five of eight LC/A MB patients (62.5%), and two of three patients (66.67%) with EP GIII. The time to progression in these patients was 2–20 months from excision (mean: 10.48 \pm 4.820 months).

Ten patients (23.3%) died after variable periods of follow up, all had disease progression (one had tumor recurrence and metastasis, and nine had metastasis). Five of the eight patients with LC/A MB (62.5%), one of the three patients with EP GIII (33.3%), three of the 14 patients with CMB (21.4%), and one of the five patients with DNMB (20%) died. No patients with

PA GI, AS GII, and EP GII subtype died during the follow up. Time to death was 2–20 (mean: 11.15 ± 4.630) months for all MB patients and 3– 18 (mean: 11.33 ± 5.203) months for ependymoma patients.

Univariate survival analysis

The estimated 2-year PFS and OS, using Kaplan-Meier statistics, for all histopathological types are listed in Table III.

Patients with CMB exhibited better PFS and OS than patients with DNMB (2-year PFS 66.0% vs. 60.0%), and both CMB and DNMB patients had better PFS than LC/A MB and EP GIII patients, who had the poorest prognosis (2-year PFS 33.3%, p=0.038).

The estimated 2-year PFS and OS for all patients were 68.1% \pm 7.4% (15.632 months) and 74.3% \pm 7.1% (16.755 months), respectively (Figure 1A, 2A). CSF seeding / metastasis (p<0.001), HR group of MB (p=0.005), subtotal resection of tumor (STR) (p=0.020), and LC/A MB and anaplastic ependymoma G III subtypes (p=0.038) all had significant negative impact on PFS and OS, i.e., they are negative predictors (Figure 1B-E; 2B-E). Age, gender, and V4 floor infiltration had no impact on outcome (Figure 1F, 2F).

Multivariate survival analysis

Cox multivariate regression was used to determine the effect of important risk factors on OS and PFS. Subtotal resection (p=0.036) was an independent negative predictor, whereas age, gender, brain stem involvement, metastasis, tumor size, and pathology had no predictive value.

DISCUSSION

The anatomy of the cerebellomedullary fissure was first depicted in 1922 by Matsushima et al. who suggested it as a potential route for approaching the fourth ventricle without neural tissue injury (16).

In our series of 44 patients, 37 patients (84.1%) had GTR, seven patients (15.9%) had STR. STR was largely due to brainstem involvement. Generally, we had no problem with tumor size or location in the superior part of the ventricle. GTR was 88.9% in Tomasello et al.'s case series (25,26), and 82.0% in Han et al.'s series (10).

In our study, we found that larger tumors tend to extend and thin the inferior medullary velum and tela choroidea, in this way encouraging a less demanding and more extensive access to the tumor that provides early access to the floor of the ventricle. The same was also reported by Tomasello et al. (25,26).

In our series, brainstem involvement was a major factor associated with STR (p=0.008) in the 24 patients (54.5%) whose tumors were invading or attached to the brainstem. GTR, meaning a complete resection or leaving a thin layer of tumor on the brain stem to avoid brainstem injury, could be achieved in 17 (70.8%) of these patients, whereas the other seven patients (29.2%) had STR. However, this problem may also occur when utilizing the transvermian route. Our results

imply that tumor adherence to the brainstem is not an absolute contraindication for total tumor resection, which is consistent with Winkler et al.'s report (28).

Tomasello et al. also reported that large tumors and tumors extending to the upper third of the fourth ventricle are more surgically challenging and increase the risk of both STR and postoperative hydrocephalus (shunt dependency) (25,26). These difficulties may also be encountered when choosing the transvermian route, but Tanriover et al. reported that the transvermian route provides a wider working angle to the rostral part of the fourth ventricle than the telovelar approach (24). Opposed to that, we did not find any problem with GTR of tumors extending to the rostral third of the fourth ventricle, because we routinely remove the posterior arch of the atlas, which provides a wider working angle. This finding is supported by Deshmukh et al.'s conclusion that the telovelar route together with removing of the posterior arch of the atlas results in a wider working angle to the upper third

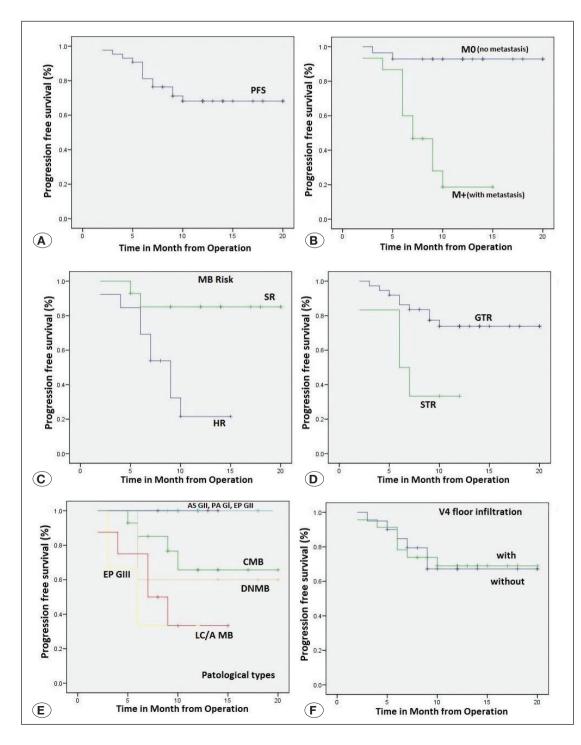


Figure 1: Kaplan-Meier estimation for PFS based on clinical factors: A) PFS in all cases; B) CSF seeding of tumor (M0 versus M+); C) HR group of MB versus SR group; D) GTR versus STR; E) Pathological subtypes; F) Tumor location with V4 floor infiltration versus without infiltration.

of the ventricle than the angle offered by the transvermian approach (4). Excision of parts of the cerebellar tonsils has been proposed as a solution to the limitations of the telovelar approach whenever the tumor is expansive or extends cranially and laterally (14). However, we believe that resection of the tonsils is not important in these cases.

We emphasize that the telovelar approach gives the advantage of early visualization and dissection of the interface between the tumor and the fourth ventricle floor, unlike the transvermian approach to the tumor from the back.

In our study, the incidence of CM was 13.6%. It had been reported that the rate of CM can be as high as 30.0% with the telovelar approach (10.29).

Gok et al., Han et al., and Matsushima et al. reported that the cases with CM all had undergone bilateral telovelar opening, whereas the occurrence of CM was improbable with unilateral

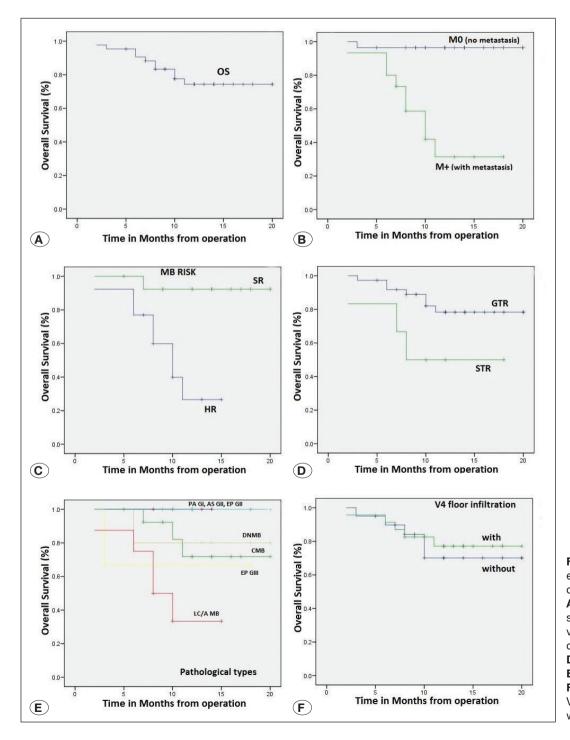


Figure 2: Kaplan-Meier estimation for OS based on clinical factors: A) OS in all cases; B) CSF seeding of tumor (M0 versus M+); C) HR group of MB versus SR group; D) GTR versus STR; E) Pathological subtypes; F) Tumor location with V4 floor infiltration versus without infiltration.

telovelar opening (9,10,17). Contrary to that, Tomasello et al. used bilateral fissure opening and encountered no case of CM (25,26). They proposed that the lack of CM in their study could be explained by the relatively higher mean age of their patients. They also suggested that cerebellar dysfunction may be avoided by wider fissure opening and limited cerebellar retraction. In our study, we used bilateral fissure opening and gentle retraction in almost all cases, but we had six cases (13.6%) of CM. We can therefore only speculate on the cause of CM in these patients. It may be related to the low mean age of our patients (6.05 years) and/or the transient post-operative edema of the cerebellum and dentate nuclei as a result of surgical manipulation or cerebellar retraction. This was also reported by Wells et al., who noticed a significant edema of the cerebellum and the middle and superior cerebellar peduncles in the postoperative imaging of patients with CM (27). In our study, CM occurred 48-96 hours post-surgery, and recovery was associated with residual dysarthria, which was previously reported and referred to as syndrome of CM with subsequent dysarthria (5). In our study, all six patients who had CM had a pathology of MB, and we therefore confirm that MB is a risk factor for CM. This is consistent with other findings reported in the literature (15,18,20,22).

In our study, we noticed a significantly higher risk of bulbar paralysis in patients who had undergone STR than in those undergone GTR (p=0.034), which may be due to brainstem invasion rather than to STR itself.

In our series, the estimated 2-year PFS and OS of all patients were $68.1\% \pm 7.4\%$ (15.632 months) and $74.3\% \pm 7.1\%$ (16.755 months), respectively. This is relatively better than Jiang et al. who reported their 5-year PFS and OS as 47.1% ± 4.6% and $54.6\% \pm 4.6\%$, respectively (13).

We found that PFS and OS were better in patients who had GTR (p=0.020), patients without CSF metastasis (p<0.001), and in the SR group of MB patients (p=0.005). This was also reported in other series (3,7,13,23).

In our series, patients with pilocytic astrocytoma GI, astrocytoma GII, and ependymoma GII had no progression during the follow-up period and the overall best outcome. Patients with CMB exhibited a better PFS and OS than DNMB, and both CMB and DNMB had a statistically significant better PFS than LC/A MB and EP GIII patients, who had the worst prognosis (p=0.038). This is consistent with the results of other clinical series (6,8), which stated that MB is associated with the worst prognosis among posterior fossa tumors. Jiang et al. in their series of CMB, DNMB, and LC/A MB found a 5-year PFS of 51.4%, 50.0%, and 9.1%, respectively (13).

The early outcome of patients is directly related to the surgical procedure and postoperative sequelae, whereas long-term outcome depends on several factors including diseaserelated morbidity and delayed effects of adjuvant therapies as described in other reports (12).

Recently, MB has been classified into four molecular types: SHH, WNT, type C/3, and type D/4 (19). Also, posterior fossa ependymoma has been classified into three molecular types: sub-ependymoma (PF-SE), PF-EPN-A, and PFEPN-B (1). Despite being similar histologically, all these molecular subgroups show a distinct behavior and prognosis, and we highly recommend performing a prospective study distinguishing between these molecular subgroups to draw definitive conclusions about their implications on outcomes.

■ CONCLUSION

The telovelar approach provides a number of merits and results in lower complication rates than the transvermian approach. It should therefore be the standard approach for fourth ventricle lesions. Early exposure of the interface between the tumor and the fourth ventricle floor, when possible, favors safer tumor resection. Whenever there is attachment of the tumor to the ventricle floor, safe resection leaving a thin layer of tumor on the brainstem is recommended to avoid brainstem injury. Wide opening of the cerebellomedullary space guarantees excellent exposure of the fourth ventricle cavity including lateral and superolateral recesses and obviates the need for excessive tonsillar retraction. This technique results in a lower risk of CM and other cerebellar dysfunctions. We recommend routine removal of the posterior arch of the atlas to gain a wider working angle, particularly to the deep rostral part of the ventricle. Tumor metastasis (CSF seeding), STR, HR group medulloblastoma, LC/A MB, and anaplastic ependymoma GIII are statistically significant negative prognostic factors.

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