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ORIGINAL ARTICLE

Hydrocephalus in patients with vestibular schwannoma

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Abstract

Background Hydrocephalus (HC) is common in patients with vestibular schwannoma (VS). This can be managed with a cerebrospinal fluid (CSF) diversion procedure prior to VS resection or with VS resection, keeping CSF diversion in reserve unless required postoperatively. No clear consensus exists as to which approach is superior. This study identifies factors predictive of the development of HC, and analyses outcomes for those managed with primary CSF resection versus tumour resection.

Methods Single-centre retrospective cohort study of 204 consecutive adult patients with a unilateral VS from May 2009 to June 2021. Data was collected on patient and tumour demographics, management, and outcome.

Results 204 patients, with a mean age at presentation of 59.5 (21–83), with 50% female, and a mean follow-up of 7.5 years (1.8–13.9) were included. 119 were managed conservatively, 36 with stereotactic radiosurgery only, and 49 with surgery. 30 (15%) patients had radiological HC, of which 23 (77%) were obstructive, and 7 (23%) were communicating. Maximum intracranial tumour diameter and Koos grade were higher in patients with HC. Of the patients with HC the majority (20, 67%) were managed initially with CSF diversion, with 12 patients undergoing subsequent tumour resection, and three patients avoiding primary resection. Nine (30%) were managed with primary surgical resection, of whom three required subsequent CSF diversion. Complication rates and Modified Rankin Scale (MRS) were comparable or lower in the CSF diversion group (8%, MRS $\leq 2 = 83\%$), versus the primary resection group (67%, MRS $\leq 2 = 67\%$), and the primary surgical resection without HC group (25%, MRS $\leq 2 = 86\%$).

Conclusions CSF diversion prior to tumour resection is a safe and acceptable strategy compared to primary VS resection, with improved outcomes and reduced surgical complications. Randomized studies and national databases are needed to determine the long-term outcomes of patients treated with CSF diversion versus primary resection.

Keywords Vestibular schwannoma · Hydrocephalus · CSF diversion · Surgery · Ventriculoperitoneal shunt

Abbreviations and acronyms

VS	Vestibular Schwannoma
CPA	Cerebellopontine Angle
CSF	Cerebrospinal Fluid
ICTD	Intracranial tumour diameter
LD	lumbar drain (LD)
EVD	external ventricular drain
VPS	ventriculoperitoneal shunt
HB	House Brackmann facial nerve (grade)
MRS	Modified Rankin Scale
MDT	Multidisciplinary team

Introduction

Vestibular schwannomas (VS) are benign tumours of the vestibular branch of the eighth cranial nerve. They arise from myelin producing Schwann cells, usually within the internal acoustic meatus where they may produce symptoms of high frequency hearing loss, tinnitus and disequilibrium. Over time they may expand into the cerebellopontine angle (CPA), and can compress structures including the trigeminal nerve, facial nerve, cerebellum and brainstem [9]. Large tumours can compress the fourth ventricle, which can impair cerebrospinal fluid (CSF) flow, causing obstructive hydrocephalus (HC) with associated symptoms of headache, nausea, and reduced consciousness. An alternative causative mechanism is that the VS can be associated with communicating hydrocephalus. This is thought to be due to a combination of CSF malabsorption at the

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arachnoid granulations secondary to micro-haemorrhages and increased protein concentration [3], and decreased intracranial compliance due to adhesions within the subarachnoid space [6, 16].

The management of VS is multimodal, largely determined by clinical presentation, the tumour size, the natural history of tumour growth and hearing function. Small VS that produce minimal symptoms are often monitored with clinical and radiological surveillance. Those that grow and become symptomatic can be managed with stereotactic radiosurgery or surgical resection. In contrast, larger tumours with more severe symptoms may be managed by primary surgical excision, with follow up surveillance and radiosurgery for residual tumour growth [5, 9]. To determine the size of VS, one usually refers to the maximum intracranial tumour diameter (ICTD) and the Koos stage [2, 17] (Table 1).

Further consideration is given to HC in association with VS, the prevalence of which varies from 1.2 to 42% [5] between studies. Several studies identified risk factors for the development of HC including larger tumour size [1, 6], older age of HC onset [4], longer disease duration [12], and higher protein concentration in CSF [6]. HC in association with VS can be managed by initial cerebrospinal fluid (CSF) diversion (by way of an external ventricular drain (EVD) or permanent ventriculoperitoneal shunt (VPS)) followed by definitive VS resection if indicated. Alternatively, cases can be managed by primary surgical resection, which results in resolution of HC in >85% of cases [5]. There is no clear consensus about which strategy is optimal and decisions depend on patient and tumour characteristics, patient choice, presentation, and surgeon preference.

This study looks at a large cohort of VS patients, to identify risk factors predictive of development of HC. It analyses the management and outcomes of both HC and non-HC patients, and specifically for those with HC, it compares those managed with primary CSF diversion versus tumour resection.

Table 1 Koos grading of VS

Grade	Definition
I	Intracanalicular tumour only
II	Tumour protrudes into CPA but does not touch brainstem
III	Tumour touches brainstem but does not displace brainstem
IV	Tumour displaces brainstem

Methods

A retrospective observational study of 204 consecutive patients with unilateral VS presenting to Queens Hospital, RomfordUK, between 14/05/2009 and 16/06/2021. The study included all adult patients, over the age of 18, with a unilateral VS identified on magnetic resonance imaging (MRI). Data collected included patient and tumour characteristics, management, complications, and outcome. This included the House Brackman grade before and after resection and the level of neurological disability at last follow-up on the Modified Rankin Scale (MRS) [13] dichotomised into those who were functionally independent (MRS score 0–2), or not (MRS 3–6). Management decisions were made by a designated skull base Multidisciplinary Team (MDT), with input from at least two consultant skull base surgeons and a radiologist. Radiological characteristics included presence of hydrocephalus, presentation and maximal ICTD and Koos grade, determined by a consultant radiologist or neurosurgeon. ICTD refers to the extracanalicular portion of the tumour along the petrous apex—for intracanalicular tumours the maximum diameter was noted. Presentation ICTD and Koos grade refer to these statistics measured on the first MRI scan that showed presence of a VS, whereas maximal ICTD and Koos grade refers to the highest Koos or ICTD measured on any MRI. Presentation and maximal Koos can be the same in cases of non-growing VS or where intervention occurred before subsequent imaging [10]. Simple descriptive statistics: 2×2 Chi-squared test with 1 degree of freedom, and 2-tailed Mann-Whitney *U* tests with level of significance defined as $p<0.05$ were used for data analysis.

Results

210 patients presented to the unit between May 2009 and June 2021, with a mean follow-up of 7.5 years (range 1.8–13.9 years). Of these, six patients had insufficient imaging and documentation available, so were excluded from the analysis.

Of the included 204 patients, 30 (15%) had HC on imaging; 23 (77%) patients had obstructive HC whilst the remaining 7 (23%) had communicating HC. Distinction between obstructive and communicating HC was made on the basis of reports by consultant radiologists.

Patients with HC had significantly larger tumours, with higher KOOS grade, than those without HC, on both the index scan and their scan identifying the maximal recorded ICTD (which is the ICTD recorded prior to intervention in operated cases). Patients with obstructive HC had

significantly larger tumours that those with communicating HC on their index scan. Similarly, patients with obstructive HC tended to have higher KOOS grade. There was no significant difference between the HC and non-HC group, or obstructive versus communicating HC in patient age laterality of tumour, or gender. These results are shown in Table 2.

Of the 30 patients identified with HC, 29 were managed surgically with either CSF diversion, VS resection, or both. One patient was not suitable for intervention due to co-morbidities. Of the 29 managed surgically, nine (31%) were managed with primary tumour resection, of which three subsequently required CSF diversion. 20 (69%) were managed initially with CSF diversion, 19 received a VPS and one an EVD. 12 of these 20 (60%) went on to have tumour resection, after a mean of 244 days (range 2–925 days). In total 21 patients with HC (70%) underwent tumour resection.

The remaining eight patients that only had CSF diversion were a heterogenous group: one tumour showed rapid spontaneous regression after VPS; two patients had resolution of symptoms, chose not to have VS resection and remain under clinical surveillance; four patients were not candidates for VS resection due to frailty and co-morbidities; and one patient developed HC after SRS—which resolved after CSF diversion and their VS regressed after the radiotherapy.

In addition to the 21 operated HC patients, 28 patients from the cohort without hydrocephalus had surgical resection of their VS. Surgical resection was mostly subtotal to preserve facial nerve function (three total, six near-total, 40 subtotal amongst all operated patients). Residual tumour was treated with stereotactic radiosurgery as required [2, 16].

Of patients with HC, those who underwent primary resection had more complications that those whose surgery was delayed until after CSF diversion. Significant complications were defined as the following: CSF leak, meningitis, post-operative bleed or intraoperative excessive bleeding, persistent postoperative CN palsy other than CN VII (measured separately) and VIII, wound infection/dehiscence or other complication requiring intervention or prolonged hospitalisation. Other measures, including rate of recurrence, time to tumour progression, functional independence (MRS score 0–2), subsequent radiotherapy, and subsequent surgery for complications of facial nerve palsy, were broadly similar between the groups (Table 3).

Outcome was measured using the Modified Rankin scale (MRS) on final follow-up. From the whole cohort of 204 patients, 84% were independent (MRS 0–2) at the end of follow-up. The distribution of MRS scores for different subgroups is shown in Graph 1.

Discussion

This study is the largest cohort study on characteristics of VS associated with HC from a UK centre, and the first globally to show improved outcomes in those managed with initial CSF diversion. In this study of 204 patients with VS, followed-up for a mean of 7.5 years, the prevalence of HC was 15%. The prevalence of HC in the existing literature ranges from 1.2 to 42% [5], with a 2021 systematic review quoting 9.3%. This systematic review looked at 273 patients with HC and VS, from 14 studies. This study is comparable with the review in terms of mean age of patients (57.4 versus 53.4) and proportion of Koos grade 3+4 (90% v 90.4% on presentation). Of note, it has a much lower proportion of communicating HC (23% versus 72%) [5]. Several studies, including this one, have shown that patients with communicating HC logically present with smaller tumours than those with obstructive HC [1, 3, 12, 16]. In this study, the ICTD was larger than in the review (39mm versus 32mm), likely due to an under representation of communicating HC [5].

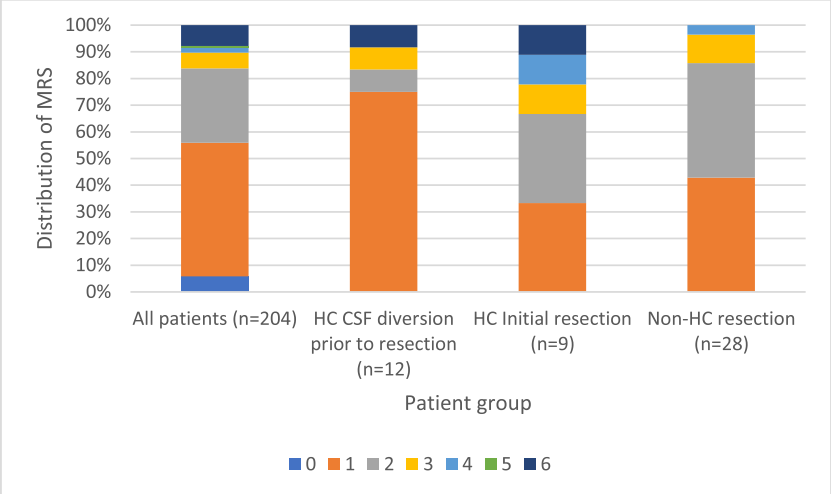
The proposed aetiology of hydrocephalus in VS is different for communicating and obstructive subtypes, though there is likely some overlap. Obstructive hydrocephalus arises from blockage of CSF drainage through the 4th ventricle and cerebral aqueduct from the tumour causing compression. Several studies have identified that a high CSF protein concentration in VS patients is associated with the

Table 2 Patient and tumour characteristics for HC vs non-HC and Communicating vs obstructive HC. Values in bold indicates significant differences at $p<0.01$, comparisons made with Chi squared and Mann-Whitney *U* test (2 tailed). Definitions of metrics used described in methods

	Non-HC	HC	*P* value	Obstructive HC	Communicating HC	*P* value
Number	174	30		23	7	
Age in years at index scan (range)	59.8 (21-83)	57.4 (25-83)	$p=0.51$	56.5 (25-83)	61.1 (46-73)	$p=0.31$
Gender (M:F)	88:86	13:17	$p=0.54$	12:11	1:6	$p=0.08$
Side (L:R)	79:95	18:12	$p=0.14$	14:9	4:3	$p=0.86$
Presentation ICTD (mm) (range)	**14.8 (3-50)**	**33.3 (10-52)**	$p<0.00001$	**36.5 (10-52)**	**25.6 (21-32)**	$p=0.0069$
Max ICTD (mm) (range)	**22.7 (3-52)**	**39.0 (21-55)**	$p<0.00001$	40.8 (32-55)	32.9 (21-53)	$p=0.151$
Presentation Koos grade (range)	**2 (1-4)**	**3.6 (2-4)**	$p<0.00001$	3.7 (2-4)	3.3 (3-4)	$p=0.08$
Maximum Koos (range)	**2.3 (1-4)**	**3.9 (3-4)**	$p<0.00001$	4 (4-4)	3.6 (3-4)	$p=0.09$

Table 3 Baseline characteristics, management, complications and outcomes of all surgically resected VS—separated into subgroups. Comparisons (using 2×2 Chi-squared test) were made between outcome measures for HC with primary CSF diversion vs non-HC, HC with primary resection vs non-HC and for HC with primary resection vs HC with primary CSF diversion. * denotes significance at $p<0.05$ on Chi-squared test. Comparisons made for all data in bold font. Facial nerve deficits requiring surgery refer to lid weights, tarsorraphy or masseteric nerve transfer. House-Brackmann grade describes facial nerve deficit at end of follow up, ranging from 0 (normal function) to 6 (total flaccid paralysis)

Characteristic	Non-HC	HC Primary resection	HC - resection after CSF diversion
Number	28	9	12
Mean age at primary resection (range)	52.3(32-84)	50.8 (26-76)	52.4 (33-73)
M:F	12:16	5:4	6:6
L:R	10:18	6:3	6:6
Translabyrinthine: Retrosigmoid	13:15	5:4	2:10
Presentation ICTD (mm) (range)	29.1 (10-50)	31.8 (10-45)	38.4 (24-52)
Maximum ICTD (mm) (range)	33.8 (17-52)	40.3 (32-49)	40.2 (24-55)
Presentation Koos grade (range)	3.3	3.4	3.8
Maximum Koos grade (range)	3.5	4	3.9
Radiotherapy	**8**	**4**	**3**
Recurrence	**6 (21%)**	**4 (44%)**	**4 (33%)**
Time to recurrence (months) (range)	**26 (15-46)**	**38 (5-80)**	**35 (20-40)**
Repeat resection	**2**	**2**	**0**
Patients with significant complication	**7 (25%)**	**6 (66%)***	**1 (8%)***
Final House Brackmann grade	1 – 20 2 – 4 3 - 2 4 - 1 5 - 1	1 – 6 2 – 2 5 - 1	1 - 8 2 - 2 3 - 1 5 - 1
Facial nerve deficit requiring surgery	**5**	**2**	**3**
Independent MRS score 0-2	**24 (86%)**	**6 (67%)**	**10 (83%)**



Graph 1 Modified Rankin Score (MRS) at end of follow-up. Separated into all patients, those with HC managed with CSF diversion, surgical resection, and non-HC surgical resection. MRS score delineated by colour.

development of communicating hydrocephalus, independent of tumour size [1, 3, 6] and that CSF pressure and protein concentration fall in tandem after VS removal [12]. It has been proposed that protein leak from the VS leads to blockage of arachnoid granulations, though meningeal adhesions and arachnoiditis likely also contribute. The blockage of arachnoid granulations may exacerbate obstructive hydrocephalus [3, 7]. Unfortunately, data on CSF protein concentration was not available for the majority of this cohort, but will be assessed prospectively.

There is no clear consensus regarding the optimal management of VS with HC [8]. A recent large cohort study advocated an individualised treatment strategy, stratifying the risk of HC related complications using severity of hydrocephalus [15] whereas the systematic review recommended primary tumour resection [5]. This is based on avoiding permanent CSF diversion as in many cases the HC resolves after primary resection (86% resolved, with only 7% requiring permanent shunting in the systematic review), and VPSs can lead to long term complications [14, 18]. The

HC resolution is due to improvement of the mechanical obstruction of the CSF drainage system, and reduction of tumour related protein, increasing reabsorption of CSF via the arachnoid granulations [3, 5, 6, 11].

In this cohort the majority of patients with HC underwent primary CSF diversion prior to consideration of surgical resection, with a small minority (10%) having no further surgical management of their VS during the follow up period. In total, 23 out of 30 (77%) patients with concurrent HC and VS were shunted, and 21 out of 30 (70%) had their tumour resected. This contrasts with the review where only 20% overall were shunted and 91% had their tumour resected [5]. In our cohort, patients managed with initial CSF diversion had similar, if not better, outcomes than those managed with primary resection, with fewer complications on subsequent tumour resection and a similar, if not higher, proportion of patients functioning independently at the end of follow up (67% versus 83% as measured by MRS).A small number of patients experienced improvement of symptoms or VS regression after CSF diversion alone, which has postponed or even avoided a larger operation for VS during the follow up period. The chance of delaying or even avoiding resection must be weighed up against risk of shunt related complications when deciding between management options. In some instances, an initial shunt may be preferable, especially when symptoms are predominantly related to hydrocephalus rather than nerve or brainstem compression.

There were several limitations to this study. This was a retrospective, single centre study, and it was not randomised. Treatment decisions were determined by the MDT, influenced by the anatomical relations of the tumour, HC versus tumour related symptoms, patient and surgeon's preference, and patient comorbidities. Although 204 was a large cohort overall, subgroups were small for comparisons and the heterogeneity of the patient group that had CSF diversion with no subsequent resection limits meaningful conclusions about avoiding resection through this approach. There was poor documentation of CSF protein. Seven and a half years of follow-up allowed assessment of tumour growth and many sequalae of surgery, but there remains a lifetime risk of growth, delayed tumour recurrence, and shunt related complications. Long term follow-up of this cohort of will continue.

Conclusion

The prevalence of radiological hydrocephalus in VS is 15%. This study demonstrates that CSF diversion prior to tumour resection is a safe and acceptable alternative or adjunct to primary VS resection. This may improve outcomes, reduce surgical complications, and perhaps in some cases avoid or delay the need for tumour resection. Further randomised studies and national databases are needed to determine the longer-term outcomes and complications of VS with HC patients treated with CSF diversion versus primary resection.

Author contributions All named authors contributed significantly to this publication, and in compliance with guidelines for Acta Neurochirurgica.

Data availability Requests for source data will be considered by authors. All patient data is anonymised.

Code availability N/A.

Declarations

Ethics approval This study is an anonymous retrospective audit; no ethics or explicit patient consent is required as per guidance from local research and development ethics team.

Consent to participate This study is an anonymous retrospective audit; no ethics or explicit patient consent is required.

Consent for publication This study is an anonymous retrospective audit; no ethics or explicit patient consent is required. All authors and interested parties have consented to publication of the manuscript.

Conflict of interest The authors declare no competing interests.

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