



# Preoperative Visual Impairment in Vestibular Schwannoma Patients: An Overlooked Entity of Developing World

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## Abstract

**Objective** Visual impairment in vestibular schwannomas is an underreported entity. The plethora of literature focuses primarily on facial and hearing preservation. This study aims to describe our experience and find the possible reasons for visual impairment.

**Materials and Methods** This is a retrospective observational study. We evaluated 114 patients with vestibular schwannoma, operated during 2015 to 2020. Eight parameters were studied—age, gender, maximum tumor size, hydrocephalus, economic status, duration of symptoms, visual loss, and ease of access to the tertiary neurosurgical facility. JASP software (version 0.14.1, Amsterdam) was used for statistical analysis.

**Results** A total of 42/114 (36.84%) patients had a varying degree of impairment. Patients with impaired vision had significantly larger tumors (4.31 vs. 4.12 cm,  $p=0.02$ ), longer duration of symptoms (19.14 vs. 16.45 months,  $p=0.02$ ), hydrocephalus ( $p=0.03$ ), and were from remote areas ( $p=0.009$ ). In stepwise logistic regression analysis, longer duration of symptoms and difficult access to neurosurgical facility stand out as decisive factors for visual impairment. The follow-up data were available for 35/42 (83.33%) cases of visual impairment. Vision improved in 14/35 (40%) of cases. Vision improved in a significantly higher number of patients when a preoperative ventriculoperitoneal shunt was placed ( $p=0.03$ ).

**Conclusion** The visual handicap occurs in a significant number of patients with vestibular schwannoma. Delayed arrival at the neurosurgical facility due to difficult access appears to be the primary factor leading to blindness. Strengthening our healthcare systems in rural areas would go a long way in the timely detection and prevention of blindness in such cases.

## Keywords

- ▶ vestibular schwannoma
- ▶ acoustic neuroma
- ▶ visual impairment

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## Introduction

Symptoms of vestibular schwannoma vary from hearing loss, tinnitus, vertigo, headache, gait ataxia, facial nerve palsy to lower cranial nerve palsy. While much focus in the literature is currently on functional preservation of cranial nerves and Gamma Knife treatment, visual impairment secondary to vestibular schwannoma had been hardly described in the published series. Neurosurgeons in developing countries commonly encounter large and giant tumors presenting with visual impairment.

Only 19.4% of the vestibular schwannomas in the United States are of large size ( $\geq 3$  cm); giant tumors ( $> 4$  cm) are even rarer (2–12%).<sup>1–3</sup> Unlike the developed world, more than 50% of vestibular schwannomas present to the developing world neurosurgeons are either large or giant.<sup>4</sup> The situation is even worse in remote regions. In India, the state of Chhattisgarh has one neurosurgeon per 3,887 km<sup>2</sup> area and 8 lac population. Most neurosurgeons and tertiary care facilities are clustered in the capital city. This distribution pattern makes access to tertiary healthcare facilities difficult for patients living in remote regions who seek neurosurgical help only when their symptoms have crippled them.

Visual impairment in vestibular schwannoma is generally attributed to raised intracranial pressure due to hydrocephalus. However, there are a subset of patients without hydrocephalus who develop visual impairment. Other hypotheses for visual impairment are also described in the literature. We intend to present our experience and describe the likely causes of visual impairment in patients with vestibular schwannomas in developing countries, like India. This study also aims to highlight this overlooked entity of the visual impairment secondary to vestibular schwannoma.

## Materials and Methods

We conducted a retrospective review of our data from January 2015 to December 2020 (6 years). There were 156 patients operated on for vestibular schwannoma. A total of 42 patients were excluded (13 patients do not have preoperative visual records, and 29 patients have refractive errors). The final analysis included 114 patients. The clinical and radiological data were retrieved from medical record department. The follow-up of the patients having preoperative visual impairment was conducted in August to September 2021, and informed consent was obtained.

A total of eight parameters were studied—age, gender, maximum tumor size in cm, presence or absence of hydro-

cephalus, economic status, duration of symptoms, visual acuity status, and ease of access to the tertiary neurosurgical facility.

The tumor was categorized into (A) tumor size less than 4 cm and (B) 4 cm or larger in maximum dimension. Hydrocephalus was defined by Evan's index more than 0.3. Economic status was categorized into below poverty line and above poverty line (APL). Visual impairment/loss was defined by World Health Organization (WHO) criteria ( $< 6/18$ ). Visual status was categorized into four groups—(A) normal vision (6/6–6/18), (B) visual impairment ( $< 6/18$ –6/60), (C) severe visual impairment ( $< 6/60$ –3/60), and (D) blind ( $< 3/60$ —no perception of light) on Snellen's chart. Visual impairment attributed to the tumor was considered when associated with concomitant fundus findings and onset of visual impairment after primary symptom of vestibular schwannoma. Access to the tertiary neurosurgical facility was dichotomized based on geographical location into easy—capital city Raipur and adjoining districts (7 districts) and difficult—remote districts (21 districts).

Statistical analysis was done using JASP software (version 0.14.1, Amsterdam). Continuous and categorical variables were expressed as mean  $\pm$  standard deviation. For categorical variables, chi-square and Fisher's exact test were used. Continuous variables were analyzed by the independent *t*-test. Stepwise logistic regression was applied to observe the influence of variables on outcome. Two-tailed *p*-values were considered statistically significant at *p*-value less than 0.05.

## Results

The final analysis included 114 patients, of which 42 (36.84%) patients had a varying degree of visual impairment (visual acuity  $< 6/18$ ). Positive fundus findings were present in 79/114 (69.29%) cases (papilloedema in 71 and secondary optic atrophy in 8 patients). Visual impairment (WHO category B) was seen in 14.91%, severe visual impairment (WHO category C) was present in 12.28% of cases, while 9.64% of patients were blind (WHO category D) (► **Table 1**). For the statistical analysis, visual functions were dichotomized into impaired vision (WHO category B, C, D) and normal vision (WHO category A).

The mean age of the total cohort was  $48.76 \pm 9.97$  (range: 22–69) years. There was no significant difference in the groups (impaired vision vs. normal vision) with respect to age, gender, and economic status (► **Table 2**).

The mean duration of symptoms was significantly higher in patients with impaired vision (19.14 vs. 16.45 months,

**Table 1** Preoperative visual status of the patients with postoperative outcome

	Preoperative status ( <i>n</i> = 114)	Postoperative improvement ( <i>n</i> = 14/35) <sup>a</sup>
Normal vision (6/6–6/18)	72 (63.15%)	–
Visual impairment ( $< 6/18$ –6/60)	17 (14.91%)	10/15 (66.67%)
Severe visual impairment ( $< 6/60$ –3/60)	14 (12.28%)	03/11 (27.27%)
Blind ( $< 3/60$ —no perception of light)	11 (9.64%)	01/09 (11.11%)

<sup>a</sup>7 patients—3 dead, 4 lost to follow-up.

**Table 2** Statistical analysis of variables for visual impairment in patients having vestibular schwannoma

Factors	Overall	Impaired vision	Normal vision	p-Value
Age (y)	48.76 ± 9.97 (22–69)	47.83 ± 9.84 (22–67)	49.30 ± 10.08 (26–69)	0.45 (NS)
Gender (M/F)	52 (45.61%): 62 (54.38%)	25 (59.52%): 17 (40.47%)	37 (51.38%): 35 (48.61%)	0.40 (NS)
DOS (mo)	17.44 ± 6.16 (6–36)	19.14 ± 5.99 (9–36)	16.45 ± 6.08 (6–30)	0.02 (S)
Maximum tumor size (cm)	4.19 ± 0.44 (3.3–5.1)	4.31 ± 0.46 (3.4–5.1)	4.12 ± 0.41 (3.3–5.0)	0.02 (S)
Tumor size category (A: < 4 cm/B: ≥ 4 cm)	40 (35.08%): 74 (64.91%)	12 (28.57%): 30 (71.42%)	28 (38.88%): 44 (61.11%)	0.26 (NS)
Economic status (BPL/APL)	89 (78.07%): 25 (21.93%)	35 (83.33%): 07 (16.66%)	54 (75%): 18 (25%)	0.30 (NS)
Hydrocephalus (yes/no)	64 (56.14%): 50 (43.86%)	29 (69.04%): 13 (30.95%)	35 (48.61%): 37 (51.38%)	0.03 (S)
Ease of access (easy/difficult)	42 (36.84%): 72 (63.15%)	9 (21.42%): 33 (78.57%)	33 (45.83%): 39 (54.16%)	0.009 (S)

Abbreviations: APL, above poverty line; BPL, below poverty line; DOS, duration of symptoms; F, female; M, male.

$p = 0.02$ ). Although the patients with impaired vision had significantly larger tumors (4.31 vs. 4.12 cm,  $p = 0.02$ ), when tumors are categorized into two groups (< 4 cm vs. ≥ 4 cm), no significant difference was observed ( $p = 0.26$ ) (► **Table 2**).

Hydrocephalus was present in 64/114 (56.14%) cases, and patients with hydrocephalus had higher chances of visual impairment ( $p = 0.03$ ). Overall, there were 42/114 (36.84%) patients from Raipur and adjoining districts, and the rest, 72/114 (63.15%), patients were from remote districts. Visual impairment was significantly higher in patients from remote districts ( $p = 0.009$ ) (► **Table 2**). A total of 28/64 (43.75%) patients underwent preoperative ventriculoperitoneal (VP) shunt placement, 22 in the impaired vision group and 6 in normal vision group.

Visual impairment was significantly associated with four factors—larger tumor size, longer duration of symptoms, presence of hydrocephalus, and difficult access to the neurosurgical facility. These factors were further analyzed in stepwise logistic regression analysis, longer duration of symptoms, and poor access to tertiary neurosurgical facility stand out as decisive factors for visual impairment. Larger tumor size and presence of hydrocephalus failed to sustain their significance (► **Table 3**).

The follow-up data were available for 35/42 (83.33%) cases of impaired vision group, three patients were dead, and four were lost to follow-up. Visual improvement was defined as an upgrade to better categories. Overall, there was a visual improvement after surgery in 14/35 (40%) of cases (66.67% in WHO category B, 27.27% in WHO category C, and 11.11% in WHO category D) (► **Table 1**). Vision improved in a significantly higher number of patients when a preoperative VP shunt was placed ( $p = 0.03$ ) (► **Table 4**).

## Discussion

### Problem Statement

The incidence of visual impairment secondary to vestibular schwannoma is challenging to determine. Among the western series, the incidence ranges from 1 to 10%, while in developing world series, it varies from 6.2 to 57.5% (► **Table 5**).<sup>3–10</sup> Actual incidences may differ due to under-reporting. In the Western world, patients seek medical attention early at the onset of symptoms; therefore, small tumors are detected early.<sup>11</sup> In a large European series of 1,865 patients, the mean tumor size was 2.2 cm in the late 1980s,<sup>12</sup> while in a contemporary Indian series of 510

**Table 3** Stepwise logistic regression analysis of studied variable: dependent variable—visual loss (present/absent), covariates—age, tumor size, DOS, factors—gender, tumor size category, economic status (BPL/APL), HCP (present/absent), ease of access (easy/difficult)

Factors	Odds ratio (OR)	Z-score	p-Value
Age	0.98	−0.65	0.51 (NS)
Gender (male)	0.72	−0.74	0.45 (NS)
Duration of symptoms	1.07	2.15	0.03 (S)
Maximum tumor size	1.45	0.37	0.70 (NS)
Tumor size category (> 4 cm)	0.55	−1.02	0.30 (NS)
Economic status (BPL)	1.19	0.32	0.74 (NS)
Hydrocephalus (present)	1.52	0.88	0.37 (NS)
Ease of access (difficult)	3.41	2.62	0.009 (S)

Abbreviations: APL, above poverty line; BPL, below poverty line; DOS, duration of symptoms; HCP, hydrocephalus; NS, nonsignificant; S, significant. Intercept included; null model applied.

**Table 4** Cross-tabulation of postoperative visual outcomes of the patient with respect to the VP shunt placement

	Vision improved	Vision not improved	p-Value
VP shunt placed	12	10	0.03 (S) Fisher's exact test
VP shunt not placed	2	11	

Abbreviation: VP, ventriculoperitoneal.

patients over 43 years, most of the tumors were large as patients here seek medical help only when sufficiently disabled.<sup>13</sup> This Indian series was categorized into three eras. Most of the patients were blind at presentation in the first decade, and 16% were blind in the middle era. Although, after the availability of the computed tomography (CT) scan (CT era), smaller tumors were picked up earlier, more than 50% had papilledema.<sup>13</sup> This report highlights the availability of neurosurgical facilities and the importance of imaging that is still a distant dream in rural areas, which comprises nearly two-third of the Indian population.<sup>14</sup> In our series, more than 50% of patients had papilloedema at presentation. It seems not much has changed in the rural and tribal India of the twenty-first century; we still get nearly one-third (36.84%) of such cases with visual impairment.

The referral pattern and presentation to the tertiary neurosurgical facility are an area of concern. It is unfortunate but true that patients with hearing loss in rural areas either self-medicate or get treated by nonregistered medical practitioners. Similarly, tinnitus and vertigo do not get adequately evaluated. Visual impairment often gets attributed to anterior chamber disease, mostly cataracts in elderly patients. These patients do not have easy access to ophthalmologists and otologists with adequate diagnostic tools in the vicinity. As long as the patient has one normal ear and visual impairment that does not significantly disable day-to-day activities, symptoms are ignored, the tumor keeps growing, and intracranial pressure keeps rising. These argu-

ments are in concordance with the observations highlighted in one Indian study.<sup>15</sup> Developing world's neurosurgeons encounter and operate mostly large or giant tumors, with the outcomes destined poor, compared with the western literature. The results of our study also highlight the significantly higher incidence of visual loss in patients from remote areas and with a longer duration of symptoms.

According to the estimates of the financial express, nearly 22% of the Indian population is below the poverty line. This is even worse in Chhattisgarh, where it is 45%.<sup>16</sup> In our study, we could not find an association between visual impairment and economic status. The likely reason is that our institute is the only state government-run tertiary care neurosurgical center that mostly provides treatment covered by government schemes; thus, practically all patients belong to the poor economic group.

It is apparent that the neurosurgeons of the developed world might not have significant cases with visual impairment due to smaller tumors, early presentation, early detection, and ease of access to a proper facility. The situation in developing world, on the contrary, is otherwise, and the neurosurgeons treat a higher proportion of large and giant tumors. Larger tumors have higher chances of having concomitant hydrocephalus and thereby a higher number of visually handicapped patients. Reporting of visual impairment gets outshined by the priority given to facial and hearing preservation efforts. We believe almost all neurosurgeons of developing world must have encountered patients of vestibular schwannomas with varying degrees of visual loss. Here, we emphasize to essentially include complete ophthalmological assessment in the preoperative workup and its evaluation postoperatively.

## Reasons for Visual Loss

### Tumor Size

We evaluated the tumor size in relation to visual loss. The mean tumor size was significantly larger in patients having

**Table 5** Review of the literature of developing and developed world describing visual impairment in patients of vestibular schwannoma

Author	n	Tumor size range (mm)	Mean tumor size (mm)	Hydrocephalus	Papilloedema	Visual impairment	Visual improvement
<b>Studies from the developing world</b>							
Jain et al 2005 <sup>4</sup>	259	11-> 40	-	-	116 (44.78%)	36 (13.89%)	-
Kumar et al 2013 <sup>5</sup>	40	> 30	-	22 (55%)	32 (80%)	23 (57.5%)	-
Huang et al 2013 <sup>10</sup>	1,009	T3-T4	38	-	-	82 (8.1%)	-
Nair et al 2016 <sup>6</sup>	64	27-66	41	40 (62.5%)	20 (31.2%)	04 (6.2%)	-
Turel et al 2016 <sup>7</sup>	179	40-67	-	155 (86.59%)	128 (71.50%)	48 (26.81%)	-
Present study	114	33-51	41.9	64 (56.14%)	79 (69.29%)	42 (36.84%)	14/35 (40%)
<b>Studies from the developed world</b>							
van Meter et al 1983 <sup>8</sup>	100	5-65	-	-	8 (8%)	02 (2%)	-
Matthies and Samii 1997 <sup>9</sup>	1,000	11-45	33	-	-	1-3%	-
Samii et al 2010 <sup>3</sup>	50	40-65	44	16 (43.2%)	13 (26%)	05 (10%)	-

visual loss (4.31 vs. 4.12 cm). When the tumors were grouped into large and giant size categories, the risk of visual loss was not different. This finding is in concordance with other studies. Huang et al in their study of 1,009 vestibular schwannomas reported no difference in visual loss between T3 and T4 categories.<sup>10</sup> Similarly, no difference was observed between the tumor size less than 5 cm and more than or equal to 5 cm by Turel et al.<sup>7</sup> van Meter et al observed that in patients with visual findings, tumor size was at least 4 to 4.5 cm.<sup>8</sup> Intracranial pressure-related symptoms have been found significantly higher in tumor size more than 4 cm.<sup>17</sup> From these reports, one may conclude that visual impairment sets in after a certain tumor size (likely 4 cm) and further growth does not change the visual status statistically.

#### Hydrocephalus/Increased Intracranial Pressure

Large and giant tumors compress the fourth ventricle leading to obstructive hydrocephalus. Hydrocephalus is seen even with smaller tumors suggesting communicating etiology.<sup>18</sup> In a series of 167 patients of Sammi et al, 2.4% of patients had hydrocephalus with a mean tumor size of 2.3 cm.<sup>3</sup> Visual impairment and papilloedema may occur even without ventriculomegaly. Matos et al reported a case of optic disc edema without hydrocephalus in a 3 cm tumor.<sup>19</sup> Papilloedema without hydrocephalus is also described in an NF 2 case.<sup>20</sup> Similar observations were reported in other studies.<sup>21</sup> A giant tumor may present with papilloedema and visual impairment without hydrocephalus.<sup>22</sup> These reports suggest that hydrocephalus is not mandatory for the development of papilloedema in vestibular schwannoma. As long as any cerebrospinal fluid (CSF) cleft persists inside the fourth ventricle, CSF circulation remains intact.

Our univariate analysis, our results suggest that larger tumor size, hydrocephalus, longer duration of symptoms, and poor ease of access to tertiary facility are the factors responsible for visual loss. However, on a multivariate logistic regression analysis, longer duration of symptoms and poor ease of access were main decisive factors. These findings reflect that larger tumor size and hydrocephalus are resultant of delayed presentation, and by improving ease of access, we can prevent visual impairment.

#### CSF Hyperproteinorrachia

Kumar et al demonstrated significantly higher protein values from the CSF of cisterna magna in vestibular schwannoma patients, which correlated significantly with visual impairment. In logistic regression analysis, duration of symptoms, papilloedema, hydrocephalus, and tumor volume was not significant for visual loss.<sup>5</sup> Shedding of protein through the tumor capsule was the probable cause. Protein concentration in ventricular CSF was also higher in their study. Bloch et al postulated that chronic inflammation, abnormal blood-brain barrier, or repeated subarachnoid hemorrhage from tumor surface vessels might lead to an increase in fibrinogen content of CSF, which ultimately gets converted into fibrin at the level of arachnoid granulations.<sup>23</sup> There is another possibility that vestibular schwannoma may produce local cis-

ternal and ventricular stasis of CSF flow, which leads to an increase in the protein levels, as seen in Froin's syndrome. Higher protein contents in CSF may also produce coagulum, which may occlude CSF flow across arachnoid granulations, resulting in an early increase in intracranial pressure without early radiological hydrocephalus.<sup>23-25</sup> Fukuda et al, in their study, found high CSF protein as a main responsible factor for hydrocephalus, and as the tumor grows, CSF protein level rises.<sup>26</sup> This cause and effect relationship is proven by the work of Miyakoshi et al, which demonstrated normalization of CSF protein contents and pressure after tumor removal.<sup>18</sup>

#### Gray Matter Alterations

Long-term hearing impairment may produce gray matter structural alterations even outside the auditory cortex. Wang et al, in their volumetric evaluation by MRI of 42 patients with unilateral hearing loss due to vestibular schwannoma, found decreased gray matter volume in the calcarine cortex.<sup>27</sup> Functional connectivity in the visual cortex gets altered in patients with unilateral hearing loss.<sup>28,29</sup> These findings suggest that visual impairment may occur even in the absence of raised intracranial pressure.

#### Prognosis

We could not find any literature on the prognosis of visual impairment after the primary tumor has been dealt with. In our study, 40% of patients had improvement in vision. Preoperative VP shunt placement helped improve vision. Patients with visual impairment need thorough ophthalmological assessments to rule out correctable causes of blindness, like anterior chamber disease. Once there is evidence of increased intracranial pressure on imaging, either in the form of hydrocephalus or tortuous and dilated intraorbital perioptic CSF spaces, early surgery is indicated. In the circumstances of heavy caseload like in a government hospital, at least CSF diversion procedures should be done while awaiting definitive surgery.<sup>7</sup> We preoperatively performed a CSF diversion procedure in 28/64 (43.75%) of patients with hydrocephalus. Jefferis et al have successfully demonstrated the benefit of optic nerve sheath fenestration in three patients of visual impairment secondary to vestibular schwannoma.<sup>30</sup>

#### Quality of Life

There are numerous studies on quality of life in patients of vestibular schwannoma.<sup>31-37</sup> The quality-of-life assessment consists of an evaluation of the patient's physical health, psychological state, and level of independence, social relationships, and relationships to their own environment. None of the published studies included visual impairment as a parameter for quality-of-life assessment.<sup>38</sup> The visually handicapped patients remain dissatisfied and live miserable lives. Once the primary devil, "the tumor," has been dealt with, the focus of these patients shifts to functional independence, and they often leave neurosurgeons baffled with the questions such as: "Why did you operate to give me a blind life?"<sup>13</sup> The surgery in these unfortunate patients does save lives, but the survivors live with the agony of deafness,



imbalance, and facial weakness; blindness adds salt to their wounds.

### Recommendations

It is evident from the present study that socioeconomic and geographical factors do play an essential role in delayed presentation. Schuz et al have suggested improving community awareness about early symptoms, specifically in the less educated and less affluent society.<sup>39</sup> We need to strengthen the people's belief in the neurosurgical services offered by the government hospitals, which, unfortunately, lag behind in self-promotion. The neurosurgeons need to break out of their shells to reach out to the people in remote areas either through continuing medical education (CMEs) or camps and need to gain the trust of their otology and ophthalmology colleagues, whom these patients consult first. Due to the lower incidence of vestibular schwannomas and poor availability of imaging facilities in remote areas, routine screening with imaging is impractical. Rupa et al have suggested evaluation with auditory brain stem response over magnetic resonance imaging as a cost-effective screening method in patients having asymmetric audio-vestibular symptoms in the hospital settings.<sup>40</sup> Early diagnosis of vestibular schwannoma is the preventive measure for visual impairment. Managing these large tumors with limited facilities requires courage and skills. It is the need of the hour for neurosurgical associations to work hand in hand with the government and create opportunities by establishing proper neurosurgical infrastructure in rural areas so that freshly graduated neurosurgeons may be motivated to provide "rural neuroscience" services.

### Strength and Limitations of the Study

The possible reasons for visual loss in posterior fossa lesions are well documented in the literature. Our results validate these observations. To the best of our knowledge, no published study had solely focused on visual impairment in vestibular schwannomas and its prognosis after treatment. Our study shows the chances of recovery in visual impairment after treatment is 40%, which could help in patient prognostication. This study highlights the existing lacunae in the healthcare delivery system in the developing countries. We believe the results of our study will raise awareness in the scientific community.

The main limitation of the study is its retrospective nature. We could not get the pretreatment fundus examination photographs to compare with the present status. In many patients, imaging was done at other center, which prevented us doing the exact volumetric analysis of tumors. For this reason, we took maximum tumor size as a parameter. In the literature, both tumor size and tumor volume had been used to assess the neurological outcomes.

### Conclusions

There are a significant number of visually handicapped patients having vestibular schwannoma. The primary reason is delayed

arrival at the tertiary neurosurgical facility due to poor ease of access in developing countries. We need to strengthen our healthcare system for early detection. The scenario of vestibular schwannomas in developing world is vastly different from that of the developed world, and patient's eyesight may still be salvaged if timely evaluated and addressed.

### List of Abbreviations

DOS, duration of symptoms; BPL, below poverty line; APL, above poverty line; WHO, World Health Organization; CT, computed tomography scan; MRI, magnetic resonance imaging; CSF, cerebro-spinal fluid; VP, ventriculo-peritoneal; HCP, hydrocephalus.

### Ethical Approval

The ethical approval was not needed and waived. The patient consented to participation.

### Informed Consent

Written informed consent was obtained from the patient to publish this manuscript and any accompanying images.

### Authors' Contributions

SK, DS, and AJ were operating surgeons; SK, DS, AJ, and MT conducted the literature search; SK, DS, and JM prepared the manuscript; SK, DS, and RS edited the final manuscript; MT, AJ, and JM performed the follow-up of the patient. All the authors have read and approved the manuscript.

### Conflict of Interest

None declared.

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### References

- 1 Babu R, Sharma R, Bagley JH, Hafez J, Friedman AH, Adamson C. Vestibular schwannomas in the modern era: epidemiology, treatment trends, and disparities in management. *J Neurosurg* 2013; 119(01):121–130
- 2 Tos M, Stangerup SE, Cayé-Thomasen P, Tos T, Thomsen J. What is the real incidence of vestibular schwannoma? *Arch Otolaryngol Head Neck Surg* 2004; 130(02):216–220
- 3 Samii M, Gerganov VM, Samii A. Functional outcome after complete surgical removal of giant vestibular schwannomas. *J Neurosurg* 2010; 112(04):860–867
- 4 Jain VK, Mehrotra N, Sahu RN, Behari S, Banerji D, Chhabra DK. Surgery of vestibular schwannomas: an institutional experience. *Neurol India* 2005; 53(01):41–45, discussion 45
- 5 Kumar B, Behari S, Jaiswal AK, Jha VK, Bettaswamy G, Singh U. Large and giant vestibular schwannomas: does cisterna magna hyperproteinorrachia influence visual status? *Acta Neurochir (Wien)* 2013; 155(01):63–69

- 6 Nair S, Baldawa SS, Gopalakrishnan CV, Menon G, Vikas V, Sudhir JB. Surgical outcome in cystic vestibular schwannomas. *Asian J Neurosurg* 2016;11(03):219–225
- 7 Turel MK, D'Souza WP, Chacko AG, Rajshekhar V. Giant vestibular schwannomas: surgical nuances influencing outcome in 179 patients. *Neurol India* 2016;64(03):478–484
- 8 van Meter WS, Younge BR, Harner SG. Ophthalmic manifestations of acoustic neurinoma. *Ophthalmology* 1983;90(08):917–922
- 9 Matthies C, Samii M. Management of 1000 vestibular schwannomas (acoustic neuromas): clinical presentation. *Neurosurgery* 1997;40(01):1–9, discussion 9–10
- 10 Huang X, Xu J, Xu M, et al. Clinical features of intracranial vestibular schwannomas. *Oncol Lett* 2013;5(01):57–62
- 11 Stangerup SE, Tos M, Thomsen J, Caye-Thomasen P. True incidence of vestibular schwannoma? *Neurosurgery* 2010;67(05):1335–1340, discussion 1340
- 12 Ben Ammar M, Piccirillo E, Topsakal V, Taibah A, Sanna M. Surgical results and technical refinements in translabyrinthine excision of vestibular schwannomas: the Gruppo Otologico experience. *Neurosurgery* 2012;70(06):1481–1491, discussion 1491
- 13 Ramamurthi B. The continuing challenge of acoustic neurinomas (1949–1993). *Br J Neurosurg* 1995;9(03):361–366
- 14 The World Bank. Rural population (% of total population) – India. Accessed June 14, 2022 from: <https://data.worldbank.org/indicator/SP.RUR.TOTL.ZS?locations=IN>
- 15 Ambett R, Rupa V, Rajshekhar V. Analysis of causes for late presentation of Indian patients with vestibular schwannoma. *J Laryngol Otol* 2009;123(05):502–508
- 16 Financial Express Available from: <https://www.financialexpress.com/economy/around-22-indians-live-below-poverty-line-chat-tisgarh-jharkhand-fare-worst/1713365/>
- 17 Bartek J Jr, Förander P, Thurin E, et al. Short-term surgical outcome for vestibular schwannoma in Sweden: a nation-wide registry study. *Front Neurol* 2019;10:43
- 18 Miyakoshi A, Kohno M, Nagata O, Sora S, Sato H. Hydrocephalus associated with vestibular schwannomas: perioperative changes in cerebrospinal fluid. *Acta Neurochir (Wien)* 2013;155(07):1271–1276
- 19 Matos RJ, Gil PN, Pires JM, Lopes N. Advanced vestibular schwannoma: a case of optic disc oedema without hydrocephalus. *Neuroophthalmology* 2016;40(05):222–224
- 20 Harada T, Sawamura Y, Ohashi T, et al. Severe optic disc edema without hydrocephalus in neurofibromatosis 2. *Jpn J Ophthalmol* 1998;42(05):381–384
- 21 Grainger J, Dias PS. Case report: optic disc edema without hydrocephalus in acoustic neuroma. *Skull Base* 2005;15(01):83–86, discussion 86–88
- 22 Candanedo C, Moscovici S, Kruger JM, Mizrachi CJ, Eliahou R, Spektor S. Vestibular schwannoma presenting with bilateral papilledema without hydrocephalus: case study. *Cureus* 2017;9(11):e1862
- 23 Bloch J, Vernet O, Aubé M, Villemure JG. Non-obstructive hydrocephalus associated with intracranial schwannomas: hyperproteinorrachia as an etiopathological factor? *Acta Neurochir (Wien)* 2003;145(01):73–78
- 24 Kwon SK, Kim MW. Pseudo-Froin's syndrome, xanthochromia with high protein level of cerebrospinal fluid. *Korean J Anesthesiol* 2014;67(Suppl):S58–S59
- 25 Garispe A, Naji H, Dong F, Arabian S, Neeki M. Froin's syndrome secondary to traumatic and infectious etiology. *Cureus* 2019;11(12):e6313
- 26 Fukuda M, Oishi M, Kawaguchi T, et al. Etiopathological factors related to hydrocephalus associated with vestibular schwannoma. *Neurosurgery* 2007;61(06):1186–1192, discussion 1192–1193
- 27 Wang X, Xu P, Li P, et al. Alterations in gray matter volume due to unilateral hearing loss. *Sci Rep* 2016;6:25811
- 28 Zhang Y, Mao Z, Feng S, Liu X, Zhang J, Yu X. Monaural-driven functional changes within and beyond the auditory cortical network: evidence from long-term unilateral hearing impairment. *Neuroscience* 2018;371:296–308
- 29 Chen J, Hu B, Qin P, et al. Altered brain activity and functional connectivity in unilateral sudden sensorineural hearing loss. *Neural Plast* 2020;2020:9460364
- 30 Jefferis JM, Raof N, Carroll T, Salvi SM. Optic nerve sheath fenestration in patients with visual failure associated with vestibular schwannoma. *Br J Neurosurg* 2019;33(04):402–408
- 31 Gauden A, Weir P, Hawthorne G, Kaye A. Systematic review of quality of life in the management of vestibular schwannoma. *J Clin Neurosci* 2011;18(12):1573–1584
- 32 Carlson ML, Tveiten OV, Driscoll CL, et al. Long-term quality of life in patients with vestibular schwannoma: an international multicenter cross-sectional study comparing microsurgery, stereotactic radiosurgery, observation, and nontumor controls. *J Neurosurg* 2015;122(04):833–842
- 33 Inoue Y, Ogawa K, Kanzaki J. Quality of life of vestibular schwannoma patients after surgery. *Acta Otolaryngol* 2001;121(01):59–61
- 34 Soulier G, van Leeuwen BM, Putter H, et al. Quality of life in 807 patients with vestibular schwannoma: comparing treatment modalities. *Otolaryngol Head Neck Surg* 2017;157(01):92–98
- 35 Wagner JN, Glaser M, Wowra B, et al. Vestibular function and quality of life in vestibular schwannoma: does size matter? *Front Neurol* 2011;2:55
- 36 Cheng S, Naidoo Y, da Cruz M, Dexter M. Quality of life in postoperative vestibular schwannoma patients. *Laryngoscope* 2009;119(11):2252–2257
- 37 Lloyd SK, Kasbekar AV, Baguley DM, Moffat DA. Audiovestibular factors influencing quality of life in patients with conservatively managed sporadic vestibular schwannoma. *Otol Neurotol* 2010;31(06):968–976
- 38 Starnoni D, Giammattei L, Cossu G, et al. Surgical management for large vestibular schwannomas: a systematic review, meta-analysis, and consensus statement on behalf of the EANS skull base section. *Acta Neurochir (Wien)* 2020;162(11):2595–2617
- 39 Schüz J, Steding-Jessen M, Hansen S, Stangerup SE, Cayé-Thomasen P, Johansen C. Sociodemographic factors and vestibular schwannoma: a Danish nationwide cohort study. *Neuro-oncol* 2010;12(12):1291–1299
- 40 Rupa V, Job A, George M, Rajshekhar V. Cost-effective initial screening for vestibular schwannoma: auditory brainstem response or magnetic resonance imaging? *Otolaryngol Head Neck Surg* 2003;128(06):823–828