

Evaluation of the Petro-occipital Transsigmoid Approach for Resection of Non-vascular lesions of the Jugular Foramen

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BACKGROUND: Lesions affecting the jugular foramen (JF) other than paragangliomas are uncommon. Of those, schwannomas and meningiomas predominate with little data describing them in the literature.

OBJECT: To evaluate the safety and efficacy of the petro-occipital transsigmoid (POTS) approach for resection of non-vascular lesions of the jugular foramen.

METHODS: This descriptive study was conducted on 12 consecutive patients affected by various jugular foramen lesions, other than paragangliomas, who were treated by the POTS approach. Computed tomography (CT) of the brain, magnetic resonance imaging (MRI) of the brain and magnetic resonance venography (MRV) were performed for all patients. The extent of resection and clinical outcome were assessed. Patients were followed up clinically by Karnofsky performance scale score and radiologically by CT and MRI for average of 1 year.

RESULTS: A single-stage resection using the POTS approach was used for all patients. Gross total resection was achieved in 66.6%. Tumors included schwannomas (6 cases), meningiomas (3 cases), chondrosarcomas (2 cases) and plasma cell tumor (1 case). This approach allowed adequate resection of both intradural and extradural components of the tumor with hearing preservation and avoidance of facial nerve transposition. There was no mortality. Variable grades of cranial nerve palsies were encountered, but none of the patients required an adjunctive procedure such as vocal cord medialization, tracheostomy, or percutaneous gastrostomy.

CONCLUSION: The POTS approach provided adequate tumor exposure for safe resection in a single-staged procedure with access to both the intracranial (extradural and intradural) and extracranial components. It allows moderate rates of hearing preservation and good rates of facial function preservation with minimal related morbidity.

KEYWORDS: Jugular foramen, non-vascular tumors, POTS approach, skull base.

INTRODUCTION

Lesions at the jugular foramen (JF) region represent challenges to the treating physicians, as these lesions involve important neurovascular structures at the JF, cranial base, posterior fossa, and high cervical region.¹ The most common tumors at the JF are paragangliomas, lesions with significant vascularity, followed by schwannomas of the lower cranial nerves and meningiomas.² The schwannomas of the JF originate from the ganglia of the ninth and tenth cranial nerves, while JF meningiomas arise from the arachnoid lining the jugular bulb.^{3,4} Primary JF lesions are tumors originating from the JF itself, growing and invading the posterior fossa to acquire an intracranial component or reaching the upper cervical region inferiorly or growing into both to acquire a dumbbell shape. However, secondary JF tumors originate elsewhere and invade the JF, presenting with various shapes and sizes e.g., chordomas

and chondrosarcomas.⁵

Clinical presentations of JF tumors did not differ according to their types and their origins. They usually present with lower cranial nerve deficits and hearing loss as their initial complaints. The most important imaging studies for diagnosis and delineation of JF lesions include a high-resolution CT scan, MRI brain with contrast, MRV and four-vessel digital subtraction angiography (DSA).⁶

The multidisciplinary team of neurosurgeons, neurotologists, and interventional neuroradiologists for management of JF lesions lead to better preoperative strategic planning and more favorable results, with the goals of radical resection and preservation of cranial nerves and vessels. The petro-occipital approach used for resection of these lesions includes a sub-occipital craniotomy combined with a retrolabyrinthine petrosectomy with transection of the sigmoid sinus. This offers the surgeon a direct exposure of the JF with preservation of the middle and inner ear structures without the need for facial nerve transposition.⁷

In this article, we present a series of 12 patients managed in our institution by the POTS approach for primary and secondary non-vascular lesions of the JF. Clinical features,

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radiological findings, operative details and surgical complications are presented and discussed.

METHODS

This descriptive study was conducted between January 2016 and January 2020 on 12 consecutive patients with non-vascular lesions of the JF who presented to Alexandria Main University hospital for management. Tumors included schwannomas (6 cases), meningiomas (3 cases), chondrosarcomas (2 cases) and plasma cell tumor (1 case). Patients with paragangliomas affecting the JF were excluded from this study. There were 11 female patients and one male patient, and the mean age was 42.5 years.

The study was submitted to and approved by our institutional ethics committee (Ethics committee, Faculty of Medicine, Alexandria University (IRB NO: 00007555- FWA NO: 00018699, <http://www.hhs.gov/ohrp/assurances/index.html>). Written Informed consent was obtained from our participants.

Clinical Evaluation

The presenting signs and symptoms were more related to the site of the pathology rather than its type, therefore many of the cases presented with similar or overlapping symptoms caused by compression of lower cranial nerves or the brain stem. Full neurological examination, head and neck evaluation, along with otologic and audiological testing were performed. Pure tone audiometry and speech discrimination scores were recorded and compared to postoperative ones. Facial nerve function was graded according to the House-Brackman (HB) scale.⁸

Headaches or a sense of neck tightness were observed in all patients, followed by dysphagia and /or choking attacks in 11 patients, hearing loss in 5 patients, and tinnitus and vertigo in 4 patients. Four patients presented with a mass invading the middle ear, appearing behind the tympanic membrane in two patients, while the other two had a polyp in the external auditory canal with superadded infections and purulent otorrhea. Three patients presented with weakness of their facial nerve, two patients with grade 3 HB and one patient with grade 6 HB. Patients were followed up clinically using the Karnofsky performance scale score.⁹

Radiological Evaluation

High-resolution CT of the skull base/petrous bone, gadolinium-enhanced MRI of the brain, and MRV of the head and neck were performed for all cases. Four patients with schwannomas underwent four-vessel DSA with suspicion of being paragangliomas. No patient underwent preoperative embolization.

Patients with JF schwannomas had tumors with smooth lobular shapes epicentered on the JF with T1 mixed signal intensity and T2 hyperintense signals. Lesions commonly showed moderate enhancement with central hypointensity due to cystic degeneration. Smooth widening of the JF was reported in petrous bone CT. One tumor exhibited a

tri-lobed appearance due to its extension into the posterior fossa, petrous bone, and high cervical region.

Patients with JF meningiomas showed isointense to hypointense tumors on T1 weighted images and intermediate signal on T2 weighted images with homogenous enhancement after contrast administration. A “dural tail” was present in all cases. Petrous bone CT showed irregular sclerosis and thickening hyperostosis of bony margins around the JF. All had intra- tumoral calcifications.

Patients with chondrosarcomas had larger tumors with extensive infiltration of the petrous bone. They appeared irregularly shaped with hypointense signal on T1 weighted images and hyperintense signal on T2 weighted images. CT petrous bone showed extensive erosion and destruction of bone. Both cases had large intracranial extensions into the posterior fossa with compression of the brain stem and cerebellum. The patient with plasma cell tumor showed a hyperintense homogenous lesion on MRI with contrast. The lesion was centered on the right occipital condyle with secondary extension into the JF.

Surgical Procedure

All patients were placed in the supine position with the head turned to the opposite side about 45 degrees. A head fixator system (Mayfield, USA) was used in most of the cases for position stabilization with care not to occlude the contralateral jugular vein. A C-shaped incision was designed starting about 4 cm above the auricle, turning 5 cm behind the postauricular sulcus, and reaching inferiorly to the level of the C-1 vertebra or the anterior border of the sternocleidomastoid muscle.

Preservation of an inferiorly based flap from the temporalis muscle and surrounding aponeurosis was a key step for skull base reconstruction at the end of surgery. The sternomastoid muscle was retracted with identification of the internal jugular vein in the neck. An extended mastoidectomy was then performed with exposure of the sigmoid sinus till the jugular bulb, followed by retrosigmoid craniotomy. A shell of bone overlying the transverse-sigmoid sinus junction was left to allow extraluminal compression with Surgicel (Johnson & Johnson, New Brunswick, New Jersey). The presigmoid dura mater in front of the sigmoid sinus was exposed with the jugular bulb completely uncovered up to the posterior semicircular canal. Identification of the mastoid segment of the facial nerve was performed with preservation of the facial nerve in its fallopian canal with exenteration of the retrofacial mastoid air cells.

Identification and sectioning of the endolymphatic sac and duct to expose the dura from the posterior surface of the petrous bone was performed, then the infralabyrinthine cells were drilled. When the tumor invaded the middle ear and/or the external auditory canal, drilling of the posterior and inferior walls of the external auditory meatus was performed with removal of the tympanic membrane together with the ossicles except for the footplate of the stapes.

The upper part of the sigmoid sinus was closed by extraluminal and intraluminal packing using Surgicel. The most important compression was achieved extradurally using a piece of Surgicel and bone wax placed between the sinus and the overlying bone shell left at the junction with the transverse sinus. The internal jugular vein in the neck was ligated. Now, as the sigmoid sinus was closed proximally and distally, it could be opened and transected for tumor removal. In all cases the sigmoid sinus was occluded in the MRV with collateral venous drainage.

The posterior fossa dura was then opened parallel to the sigmoid sinus for further exposure of the intradural part of the tumor. This opening was connected to the presigmoid dural opening by the transection of the occluded sigmoid sinus with a horizontal incision across it traversing the medial wall of the sinus. Bleeding from the inferior petrosal sinus was controlled by gentle packing with Surgicel.

Exposure of the intradural part of the tumor required no or minimal cerebellar retraction. The 7th and 8th cranial nerves were usually identified above the lesion, while lower cranial nerves identification could be difficult especially in larger tumors requiring meticulous microsurgical dissection.

Before closure, obliteration of all opened mastoid labyrinthine cells with bone wax is mandatory. The eustachian tube opening was obliterated with a piece of muscle. At the end of the procedure, the retrosigmoid dura was closed, and the rest of the cavity was packed with abdominal fat. By re-suturing of the inferiorly based flap from the temporalis muscle and surrounding aponeurosis, the fat strips were left in place with a certain degree of compression. When drilling of the external auditory canal was performed, the external auditory meatus was sutured-closed in a blind- sac fashion and re-enforced with a layer of the musculo- periosteal flap.

Postoperative clinical outcome was assessed and the extent of resection was evaluated and classified into gross total and subtotal resections. Follow-up was performed in the form of MRI scans at 3 and 6 months, and then annually.

The study was submitted to and approved by our institutional ethics committee (Ethics committee, Faculty of Medicine, Alexandria University (IRB NO: 00007555-FWA NO: 00018699, <http://www.hhs.gov/ohrp/assurances/index.html>).

RESULTS

Twelve patients were included in the study, 11 females and 1 male. Tumors included 6 schwannomas, 3 meningiomas, 2 chondrosarcomas originating in the petrous bone with secondary erosion in the ipsilateral JF, and one plasma cell tumor that originated in the bone of the sub-occipital area and condyle and encroached extradurally on the nearby JF.

Gross total removal was achieved in 8 patients (66.6%), all 6 patients with schwannomas and 2 of the patients with meningiomas (**Table 1**). Total removal was not possible in 4 patients including 2 patients with chondrosarcomas due to extensive bony, dural, and nerve invasion and infiltration, and one patient with plasma cell tumor also due to extensive bony involvement. The last patient with subtotal meningioma resection had a large upper cervical component with good preoperative lower cranial nerves function, so incomplete resection was planned to preserve the lower cranial nerves functional outcome with the residual cervical portion to be followed up with serial MRI scans that showed stationary course.

The most frequent and life-threatening complication was a new deficit of the lower cranial nerves, that could potentially lead to aspiration pneumonia and death. Eleven patients (91.6%) had deterioration in their lower cranial functions after surgery, which was transient in all cases with none developing aspiration pneumonia. Eight patients had worsening of their hearing and word recognition after surgery (66.6%). Three patients had weakness of their facial nerve, with two presenting with an HB grade 3 function which improved after surgery to HB grade 1 function. The remaining patient presented with an HB grade 6 preoperatively due to invasion of the facial canal, which did not improve after surgery.

One of the patients with invasive chondrosarcoma had an ipsilateral complete abducent palsy that did not improve with follow-up. The other patient with large chondrosarcoma extending into the posterior fossa suffered transient hemiparesis due to compression of the ipsilateral middle cerebral peduncle. One patient with a JF schwannoma experienced a cerebrospinal fluid (CSF) leak from the wound and was managed with over suturing of the wound and placement of a lumbar drain (**Table 2**). No problems or morbidity occurred due to venous obstruction related to sigmoid sinus sectioning. There were no perioperative deaths. (**Table 3**).

Table 1: Histopathological criteria and extent of resection

Histological exam of tumor	Number of cases	Extent of resection	
		Total	Subtotal
Schwannoma	6	6	0
Meningioma	3	2	1
Chondrosarcoma	2	0	2
Plasma cell	1	0	1

Table 2: Surgical complications

Morbidity	Number
Lower cranial nerves palsy	11
Vestibulo-cochlear nerve palsy	8
Facial nerve palsy	3
Abducent nerve palsy	1
Cerebrospinal fluid leak	1
Hemiparesis	1

Table 3: Karnofsky performance scale score

Able to carry on normal activity and to work; no special care needed	12
Unable to work; able to live at home and care for most personal needs; varying amount of assistance needed	0
Unable to care for self; requires equivalent of institutional or hospital care; disease may be progressing rapidly	0

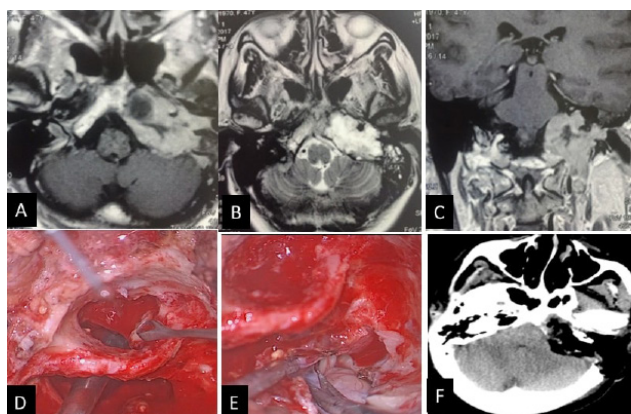
Illustrative cases (Figs. 1-4).

Fig. 1: A 47 years old patient complaining of headache, tinnitus and hearing loss on the left side with a previous history of left offensive otorrhea. Examination showed grade 3 HB facial function which developed over 3 months. Otologic examination revealed a polyp in the left external auditory canal. Pure tone audiometry showed severe mixed hearing loss. A: Axial T1 MRI shows a lobulated hyperintense mass with cystic degeneration on T1. B: Axial T2 MRI shows a lobulated mass with mixed signal encasing the left internal carotid sheath inferiorly. C: Coronal MRI with contrast shows extension into the upper cervical region. D: The facial nerve is kept protected in its fallopian canal with resection of the tumor anterior and posterior to it. E: Opening of the dura mater for resection of the small intradural component; the 7th, and 8th cranial nerves are seen entering the internal auditory canal. F: Postoperative CT images showing the extent of bony drilling and complete resection of the JF schwannoma with fat filling the surgical bed of the POTS approach.

DISCUSSION

The JF comprises a complex bony and neurovascular anatomy. Anatomical studies have been performed to describe it, but the surgical anatomy of this region remains poorly understood. A good understanding of the anatomy of the related regions, like the temporal bone, posterior fossa, and high cervical region is needed for resection of lesions in the JF.^{10,11}

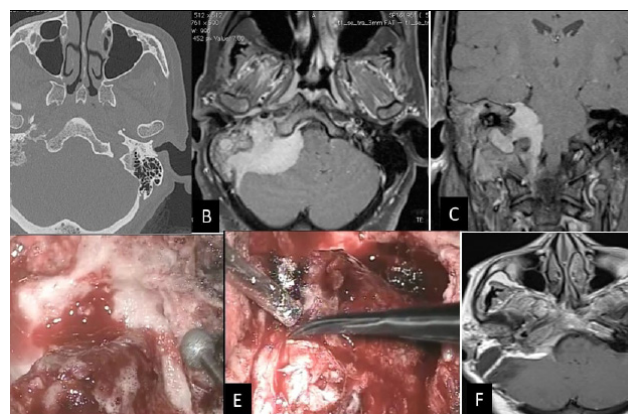


Fig. 2: A 40 years old patient presenting with right-sided tinnitus and occipital headaches for five years, and gradual hearing loss over 6 months in the right ear. A: Head CT shows hyperostosis and sclerosis of the right mastoid/petrous bone with widening of the JF, B: Axial MRI with contrast shows a sizable enhancing mass filling the JF, sigmoid sinus and jugular bulb with a dural tail along the clivus, C: Coronal MRI with contrast shows that the tumor extended well into the neck in the retropharyngeal space, D: At the end of drilling of the bone covering the sigmoid sinus, the sinus is seen full of tumor, E: Cutting of the dura mater of the posterior fossa parallel to the edge of the sigmoid sinus, followed by cutting of the sigmoid sinus using micro scissors after its coagulation, F: Postoperative axial MRI with contrast showing near-total resection of the right JF meningioma, with a small residual along the dural tail traversing the JF.

Patients with JF tumors usually present to otolaryngology surgeons as they commonly manifest with symptoms affecting hearing and swallowing. Growth of the lesion may also cause vertigo, hoarseness, paralysis of the tongue, and rarely facial paralysis or a polypoid mass in the external auditory canal. However, neurosurgeons usually see patients in more advanced stages with cerebellar manifestations and hemiparesis resulting from large intracranial components in the posterior fossa.¹⁰

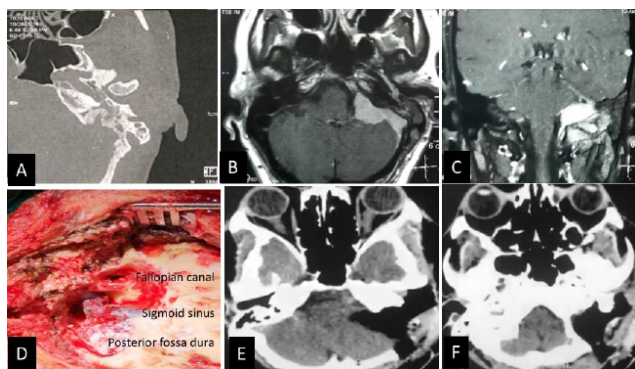


Fig 3: A 60 years old patient presenting with significant occipital headaches and repeated attacks of dysphagia. **A:** CT of the left petrous bone showing hyperostosis and sclerosis of the bone surrounding the jugular foramen, **B & C:** Axial and coronal MRI with contrast showing enhancing meningioma epicentered on the left JF with invasion of the sigmoid sinus with a dural tail, **D:** Surgical field after performing a left POTS approach; the facial nerve is left intact within the fallopian canal with complete skeletonization of the sigmoid sinus and jugular bulb. A small suboccipital craniotomy with exposure of the posterior fossa dura is performed, **E & F:** Post-operative CT showing the extent of bony removal for surgical exposure. The cavity is filled with strips of abdominal fat for the prevention of a CSF leak.

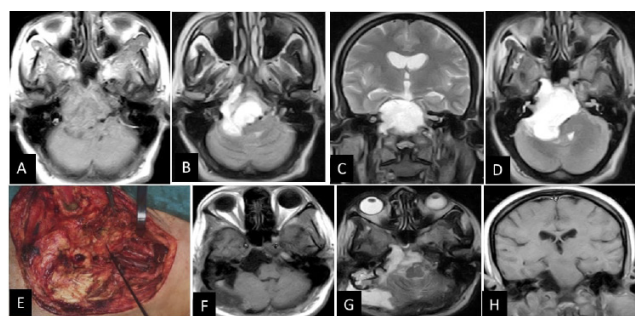


Fig 4: A 39 years old patient presenting with headache, right facial pain, and left-sided weakness. **A:** Axial T1 MRI, **B:** Axial T2 MRI, **C:** Coronal T2 MRI, **D:** Axial T2 MRI showing a large right chondrosarcoma with secondary involvement of the right JF. The tumor extends anteriorly to involve the clivus with compression of the cerebellum and brain stem, **E:** Surgical exposure of the mastoid bone and upper cervical region with dissection of the jugular vein and carotid artery. A nerve stimulator is used to identify the facial nerve before the parotid fascia. The skin of the external auditory canal is transected and the whole flap is retracted anteriorly, **F:** Axial MRI with contrast, **G:** Axial T2, **H:** Coronal MRI with contrast showing subtotal resection of the tumor with residual tumor along the right petrous carotid canal, with very good decompression of the brain stem and posterior fossa.

After exclusion of paragangliomas, non-vascular primary JF tumors in this series were most commonly schwannomas and meningiomas, but the JF was also secondarily affected by chondrosarcomas and less frequently by plasma cell tumor. In this study, JF schwannomas were the most common accounting for 6 cases. The most common presentation

was choking attacks along with headaches and decreased hearing accompanied by vertigo and imbalance. One patient with a recurrent schwannoma operated elsewhere had a complete facial weakness as well as a polypoid mass in the external auditory canal when referred to our group. Meningiomas came in second with 3 cases, with most patients reporting difficulty in swallowing and headaches as their main complaint. Two patients were found to have decreased hearing with decreased word recognition below 50%. Chondrosarcomas were encountered in 2 patients with significant intracranial components leading to significant imbalance and contralateral weakness due to cerebellar and brain stem compression. One patient had a plasma cell tumor presenting with severe occipital and neck pain and repeated choking attacks. This was like larger series of patients where schwannomas constituted 16%, followed by meningiomas 10% and then chondrosarcomas in 5% of the patients with lower percentages for rare lesions affecting the JF.^{12,13}

Radiological diagnosis of JF lesions, being rare tumors, can be commonly misleading. Half of the lesions in this cohort were initially reported as paragangliomas. One patient with a JF schwannoma underwent DSA after initially being reported as a glomus tumor before being referred to our service with the angiography expectedly not showing the extensive vascularization/blush associated with glomus tumors.

The best treatment for benign JF tumors is radical removal. Complete resection with the preservation of the cranial nerves and vessels remains a challenge in many cases.¹⁴ A multidisciplinary skull base team with experience of neurosurgeons and otolaryngological surgeons allows the best chance of radical removal with preservation of the involved structures and tackling challenges associated with resection of these lesions like deep location, cranial nerves, and vessels infiltration, bone erosion, and extensions within the posterior fossa. Adequate tumor exposure and cranial base reconstruction are necessary to avoid postoperative complications.

Utilizing the POTS approach to resect these lesions was performed in all cases. This approach was first introduced by Mann et al in 1991¹⁵ and modified by Mazzone and Sanna in 1995.⁷ This allowed adequate tumor exposure for resection with access to both intracranial and extradural tumor parts without facial nerve rerouting and the possibility of preservation of hearing. It also allowed perfect exposure of the intradural component of the tumor allowing adequate resection of tumors commonly reaching large sizes before presenting clinically in a single staged procedure.

The gross total resection of all JF schwannomas was feasible via the POTS approach. Gross total resection was less achieved in JF meningiomas, only in two patients while one patient had subtotal resection. This coincides with other reports in the literature showing the same rates for resections for schwannomas versus meningiomas.¹⁶ This might be explained by the higher tendency of meningiomas to infiltrate dura, bone, and vessels thus requiring a more

aggressive surgery. A near-total resection was achieved for JF chondrosarcomas as well as for the patient with the plasma cell tumor.

Overall, the major morbidity following this approach for JF tumors was caused by lower cranial nerve deficits leading to difficulty or worsening of swallowing. This seemed to be related to the location of the pathology itself rather than the approach. Long-term facial nerve function preservation of an HB grade I was achieved in all patients in this series except for one case with a recurrent trilobed JF schwannoma operated elsewhere who presented with a grade 6 HB facial function that did not improve after surgery. The reason for good facial function was the preservation of the facial nerve protected in its fallopian canal during drilling and meticulous microsurgical preservation of the nerve intradurally guided by intraoperative monitoring. Hearing preservation was possible in 4 of the 12 patients.

None of the patients in this series had any complications as regard sacrificing the sigmoid sinus. In addition, none of the patients required a tracheostomy or a feeding gastrostomy. Only one patient had a CSF leak from the retro-auricular incision and was managed with overrunning sutures and applying a lumbar drain.

CONCLUSION

Non-vascular tumors of the jugular foramen are rare neoplasms that still pose significant diagnostic and therapeutic difficulties in modern neurosurgery. Surgical resection of these tumors via the POTS approach provided adequate tumor exposure for resection in a single staged procedure with access to both intracranial (extradural and intradural) and extracranial components. The POTS approach achieved moderate rates of hearing preservation and good rates of facial function preservation with minimal related morbidity.

List of abbreviations

CSF: Cerebrospinal fluid.

CT: Computed tomography.

DSA: Digital subtraction angiography.

HB: House-Brackman.

JF: Jugular foramen.

MRI: Magnetic resonance imaging.

MRV: Magnetic resonance venography.

POTS: The petro-occipital transsigmoid approach.

Disclosure

The authors report no conflict of interest in the materials or methods used in this study or the findings specified in this paper.

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