

OTOLOGY

Closure of the sigmoid sinus in lateral skull base surgery

Chiusura del seno sigmoide nella chirurgia della base cranio laterale

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SUMMARY

Closure of the sigmoid-jugular complex is generally planned during various surgical procedures on the skull base, either to repair a jugular foramen lesion or as the oncological boundary of the resection. A series of 218 cases of skull base tumour surgeries was analysed in which closure of the sigmoid-jugular complex was systematically planned (bilaterally in one case) in patients treated for jugular foramen paragangliomas, squamous cell carcinomas and other temporal bone tumours. Surgery was performed via a petro-occipital trans-sigmoid approach in 61 cases, an infratemporal A in 128, *en bloc* subtotal temporal bone resections in 10 and other approaches in 20. In our experience, planned unilateral (and, in one case, bilateral) closure of the sigmoid-jugular complex had no clinical consequences. The vicarious drainage of the skull base was always assessed preoperatively, revealing no contraindications to intraoperative sinus closure. Given the scarcity of literature on this subject, the present report shows that the procedure is associated with low morbidity and helps to improve our understanding of cerebral venous discharge.

KEY WORDS: Sigmoid sinus closure • Lateral approaches • Jugular foramen surgery

RIASSUNTO

La chiusura del complesso seno sigmoide-bulbo giugulare è contemplata in alcuni approcci chirurgici laterali al basi cranio; può essere parte dell'accesso chirurgico stesso al forame giugulare, oppure risultare necessaria come margine di una resezione oncologica del basi cranio laterale. Abbiamo analizzato una casistica personale di 218 pazienti trattati chirurgicamente per patologie tumorali della base cranica, sottoposte a chiusura pianificata del complesso sigmoide-giugulare. In un caso la procedura è stata eseguita bilateralmente. Tra le patologie trattate erano presenti paragangliomi del forame giugulare, carcinomi squamo cellulari dell'osso temporale ed altri tumori dell'osso temporale. Sono stati eseguiti 61 approcci petro-occipitali, 128 approcci infratemporal di tipo A, 10 resezioni subtotali en-bloc dell'osso temporale e 20 approcci di altro tipo. Nella nostra esperienza, la chiusura pianificata unilaterale – bilaterale in un singolo caso – del complesso sigmoide-giugulare non ha mostrato conseguenze cliniche. Per ogni paziente è stata attuata una valutazione preoperatoria del sistema di drenaggio venoso intracranico, che non ha rilevato controindicazioni alla procedura di chiusura. Considerata la carenza di dati in letteratura su questo argomento, il presente lavoro offre alcuni spunti per lo studio dei meccanismi di drenaggio venoso intracerebrale in condizioni fisiologiche e patologiche e dimostra la bassa morbilità della chiusura del complesso sigmoide-giugulare.

PAROLE CHIAVE: Chiusura seno sigmoide • Approccio laterale • Chirurgia del forame giugulare

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Introduction

Experience with skull base surgical procedures has shown that intraoperative closure of the sigmoid sinus and jugular bulb (sigmoid-jugular complex; SJC) has no clinical consequences¹.

Closure of the SJC may be planned as part of surgical procedures involving the jugular foramen (using an infratemporal A, a petro-occipital trans-sigmoid, or a far lateral approach), or to meet the needs of oncological radicality, as in subtotal *en bloc* temporal bone resections.

SJC closure may also be unplanned but prove necessary during lateral skull base surgery when tearing, coagulation or trauma occur (even without any obvious lesions)

in procedures involving translabyrinthine or retrosigmoid approaches¹.

In cases of tumour involving the jugular foramen, the SJC may already be partially or completely closed by the tumour, whereas the lumen is free in other cases of closure (either unplanned or part of *en bloc* resections). In both conditions, the resulting obstruction of the venous discharge from the brain and skull base has no clinical consequences. Transient cerebral oedema has been observed in rare cases, with no associated clinical signs²⁻⁴, and serious consequences for the central nervous system are exceptional. When they do occur, they are not due the SJC closure per se, but rather to concomitant conditions (e.g. anatomical variations, lack of compensatory mechanisms, latent diseases) behind such clinical consequences.

Venous drainage from the brain has sufficient alternative routes⁵, both in physiological conditions and after closure of the SJC. The anatomical and functional aspects of cerebral venous discharge are discussed here, together with a report on our experience of planned SJC closures. The rates of unplanned SJC closure are probably underestimated because they do not give rise to functional consequences.

Materials and methods

At our tertiary referral centre, 218 patients with skull base tumours were treated surgically with planned closure of the sigmoid sinus between 1985 and 2004. SJC closure was bilateral in one case (Table I).

The series included 64 cases of various jugular foramen (JF) tumors, 144 of C class paragangliomas and 10 primary squamous cell carcinoma of the external auditory canal. The petro-occipital trans-sigmoid (POTS) approach⁶ was used in 60 patients (including one with bilateral chondrosarcoma of the JF who underwent two staged procedures with bilateral closure of the SJC), the infratemporal A (IT-A) approach in 128, other approaches in 20 and subtotal *en bloc* temporal bone resections (STBR) were performed in 10 cases⁷.

All patients were managed by the same senior surgeon using a consistent technique.

In all 219 procedures, the sigmoid sinus and jugular bulb complex was either closed as part of the surgical procedure (in 61 POTS, 128 IT-A, and 20 other approaches), or necessitated by subtotal *en bloc* bone resection (10 STBR).

The lesions originated in or near the JF (schwannoma, paraganglioma, meningioma), or grew to involve the jugular fossa (chordoma, chondrosarcoma, cholesteatoma). In all cases, the lesion extended to a variable degree into the cerebello-pontine angle (CPA), skull base bone and neck. In *en bloc* temporal bone resections (squamous cell

carcinoma of the external auditory canal and temporal bone), the SJC was free of disease but was included in the resections for the sake of oncological radicality. Diagnoses were always obtained with contrast-enhanced CT scans and, since the 1990s, with contrast-enhanced MRI and CT scans. Preoperative angiography was used to investigate venous discharge status through the sinuses and patency of the torcular herophili.

Results

Sixty-one POTS procedures (1 bilateral) were performed for 11 type C jugular foramen paragangliomas and 49 other jugular foramen tumours; 128 IT-A were performed in 113 cases of type C jugular foramen paraganglioma and in 15 patients with other JF lesions; other approaches were used for 20 type C jugular foramen paragangliomas (Table I). In all these procedures, the SJC was closed due to tumour involvement or as part of the surgical procedure.

Cases of primary squamous cell carcinoma of the external auditory canal were treated with *en bloc* STBR. The SJC complex was sacrificed because, though free of disease, it was within the oncological boundaries for the purposes of radical tumour removal.

In all cases, closure of the SJC had no clinical consequences. The case of bilateral sinus closure was a patient with bilateral chondrosarcoma of the JF who was treated with staged POTS. No anomalies came to light on preoperative venous drainage assessment, and none of the patients had any preoperative contraindications to closure of the SJC.

Discussion

Closure of the sigmoid sinus may either be planned or as part of an unintentional outcome of transpetrosal surgical procedures¹.

Table I. Case material of skull base tumours treated surgically using various approaches involving closure of the sigmoid-jugular complex.

Total cases of jugular foramen (JF) – temporal bone tumours	POTS	IT-A	Other approaches	En bloc STBR
	No. of cases	No. of cases	No. of cases	No. of cases
Schwannoma 9-11 cranial nerve	16			
Schwannoma 7,8,12 cranial nerve	4			
JF paraganglioma (first diagnosis)	6	98	17	
JF residual paraganglioma	5	15	3	
Meningioma	7			
Chordoma	4	1		
Chondrosarcoma	5	3		
Papillary adenoma	3	4		
Miscellaneous	10	7		
Primary squamous cell carcinoma of the external auditory canal				10
Total cases (N = 218)	60	128	20	10
Total procedures (N = 219)	61	128	20	10

The reason why it has no functional consequences is probably because compensatory drainage mechanisms already exist in physiological conditions, but only become apparent when the SJC is closed.

The anatomy and physiology of venous drainage from the brain and skull base involve a rich network of emissary veins connecting the vessels outside the skull with the intracranial venous sinuses⁵. This network is more evident in children and not all the emissary veins are identifiable in each individual⁸. These veins are valveless and blood can flow bidirectionally. Their function consists in equalizing intracranial pressure, and they act as safety valves at times of cerebral congestion or when the SJC becomes obstructed. Most of these veins connect various subsites of the SJC with the internal vertebral plexus. The posterior condyloid vein runs through the posterior condylar foramen in the retromastoid region, connecting the internal vertebral plexus to the lower end of the sigmoid sinus. The mastoid emissary vein passes through the mastoid canal and connects the sigmoid or transverse sinus to the postauricular or occipital vein, which joins the internal vertebral plexus.

Gilbert Breschet provided the first detailed description of the anatomy of the vertebral venous plexus in 1819⁹. It was described as a large plexiform, valveless network of vertebral veins consisting of three interconnecting divisions and spanning the entire spinal column, with connections to the cranial dural sinuses distributed in a longitudinal pattern, running parallel to and communicating with the venae cavae. It is now considered as being functionally even more important than the SJC in the cerebrospinal venous system¹⁰. It plays an important part in regulating intracranial pressure with changes in posture, and in venous outflow from the brain. In pathological conditions, it provides a potential route for the spread of tumour, infection, or emboli.

The emissary veins are routes connecting the intracranial sinuses to the vertebral venous plexus. The occipital emissary vein connects the transverse sinus with the internal vertebral plexus through the occipital vein; the parietal emissary vein connects the superior sagittal sinus to the occipital vein, and then to the internal vertebral plexus. Other emissary veins connect areas other than the SJC, e.g. the foramen ovale, sphenoid sinus, foramen cecum, foramen lacerum and clivus.

Batson et al.¹¹ studied all the venous channels and emissary veins draining the adult skull base, and found that venous outflow from the brain is via the internal jugular veins, the anterior anastomoses with the orbit and pterygoid plexuses, the emissaries through the skull and multiple channels joining the vertebral plexuses¹².

All these plexuses operate under normal physiological conditions as in coughing, sneezing, straining, and jugular compression. In some individuals, they are capable of completely taking over blood drainage after acute occlusions of the dural sinuses or jugular veins. If these col-

lateral drainage channels were to prove inadequate due to an abnormal occlusion of the major dural venous sinuses, then intrasinal venous pressure would probably increase. In some patients, the sinus is functionally already blocked by tumour, and its closure during surgery does not alter the situation existing preoperatively. Any increase in intrasinal venous pressure can be compensated providing the collateral channels are adequate in number and calibre, and capable of diverting the blood into appropriate exit channels¹².

This network of emissary veins is not always the same in every individual, and this may explain the variety of compensatory mechanisms occurring (and the corresponding clinical aspects) in the event of SJC closure.

When closure of the SJC is an involuntary intra- or post-operative event, it may be due to thrombosis of the sinus, intraoperative tearing of the sinus wall, heat-induced injury during bone drilling, or extrinsic compression¹³⁻¹⁵. Other intraoperative events, such as retractions, parenchymal damage, or injury to the venous system, may make an otherwise asymptomatic event become clinically evident. Very few cases of unplanned sigmoid sinus occlusion have reportedly had serious clinical consequences, and this has been attributed primarily to anatomical variations in the intracranial venous system^{2,4,16,17} and to individual differences in the whole collateral venous circulation network. Preoperative angiography to assess the collateral venous system is commonly recommended before taking a transpetrosal approach to the jugular foramen. When sacrifice of the sinus is planned, occlusion of the sigmoid sinus is safe and symptom-free in the case of a freely communicating torcular herophili, but closure of the sigmoid sinus may have consequences if the lesion is located at the same site in the dominant sinus, the torcular system is only partially communicating and the collateral venous discharge proves insufficient^{4,18}.

Preoperative assessments are not usually prescribed when conventional transpetrosal procedures involve the jugular foramen^{19,20}, and the unplanned sacrifice of the sigmoid sinus has never reportedly been a problem *per se*. It may be that a system of collateral drainage and emissary veins compensates for the sigmoid sinus closure. Alternatively, the fact that closure of the SJC has no adverse effects might be attributed to the slow growth of tumours affecting the jugular bulb prompting a vicarious blood discharge. The same lack of clinical consequences is seen after *en bloc* resections, however, in patients whose SJC had an unobstructed venous flow before surgery, giving the impression that alternative routes may open when the sigmoid sinus is closed. A right dominance of the sigmoid sinus has been reported in 60% of patients, a left dominance in 25% and an asymmetrical dominance in 15% of cases^{12,21}.

The concept of "dominant sinus" has always been discussed on the basis of angiographic evidence, but the ap-

parently dominant pattern has not been investigated from a functional standpoint. It is not yet clear whether it is safe to sacrifice the so-called 'dominant' sigmoid sinus. The collateral unilateral drainage of the non-dominant sinus would seem to be plentiful and compensate for the apparent reduction in venous flow¹². Our case of bilateral (albeit staged) closure of the SJC is of interest, given the lack of any adverse effects in this patient.

Sigmoid sinus thrombosis is also a silent complication of otomastoiditis and its outcome is favourable in most cases. When life-threatening sinus thrombosis is associated with cerebral oedema and symptoms of intracranial hypertension (pseudotumour cerebri), this generally coincides with inadequate collateral venous discharge¹⁸.

Symptoms of intracranial hypertension include headache, blurred vision and vomiting²². Examining the fundus oculi always reveals papilloedema, and often retinal haemorrhage. These symptoms are rarely life-threatening and usually self-limiting, and patients respond to conservative treatments such as steroids and diuretics.

Two mechanisms have been described to explain the cerebral dysfunction that accompanies cerebral venous sinus thrombosis leading to an increase in intracranial pressure⁴. Cortical vein thrombosis causes local venous hypertension and leads to vasogenic and ultimately cytotoxic oedema, ischaemia and haemorrhage. Sinus thrombosis causes intracranial hypertension by impeding CSF absorption at the arachnoid villi, which are located primarily in the superior sagittal and transverse sinuses. Thrombosis of the sinuses and the resulting increase in venous pressure hinders CSF flow into the ventricles, but this does not usually cause hydrocephalus. The intracranial pressure only increases if and when the anastomotic channels between the sinuses and elements of the cerebral venous system proximal to the site of obstruction are inadequate²³. The reason why the ventricle does not increase in size with obstructive hydrocephalus is not clear⁴.

Some conditions existing preoperatively might make it more likely for symptoms to become clinically evident, e.g. infections or chronic diseases such as collagen tissue disorders, cardiac disease, haematological abnormalities and venous abnormalities in the brain's drainage pattern. One in two patients who develop symptoms after cerebral venous thrombosis have a previously unknown prothrombotic state that may relate to anticardiolipin antibodies, deficiencies in proteins C and S, or antithrombin III, or prothrombotic gene mutations involving prothrombin 20210, factor V Leiden, or MTHFR^{18 22 24}.

There is general consensus^{2 4 16 17} that iatrogenic occlusion of the sigmoid sinus is usually uneventful, though there is a paucity of literature on experimental studies. Kanno et al.³ investigated the consequences of acute sigmoid sinus closure in monkeys. The hypothesis that sacrificing the sigmoid or transverse sinus had no consequences was confirmed experimentally by the lack of any significant increase in venous

pressure. The pressure in the contralateral transverse sinus did not change significantly when the sigmoid sinus was occluded³. This might be because blood does not always flow in the same direction and, when the sigmoid sinus is occluded, flows from the transverse sinus to the contralateral transverse sinus through the vein of Labbé and the torcular vein²⁵. Any disturbance caused by the sinus occlusion can thus be compensated by changing the direction of flow. When the transverse sinus is occluded, for instance, more blood drains through the vein of Labbé and the superior petrosal sinus to the sigmoid sinus. It is therefore essential for the vein of Labbé, the superior petrosal sinus, and the bilateral transverse sinuses to be visible on a preoperative angiogram. Otherwise sigmoid or transverse sinus ablation might raise sinus pressure, thereby causing venous congestion and brain swelling, and becoming symptomatic. These experimental findings are important, but it is important to know whether they also apply to humans. Recent reports^{3 25} and common experience suggest that accidental closure of a sigmoid sinus is generally without clinical consequences, but clinical or experimental studies on the outcome of unplanned or accidental sinus ablation in humans remain limited.

When the clinical consequences of cerebral venous thrombosis become evident, their management becomes mandatory. The limited evidence available makes the appropriate management of internal jugular and cerebral sinus thrombosis hard to establish. Most of the literature concerns single case reports or small case series, but there are no controlled studies (due to the rarity of these conditions), and so there is no broad agreement or policy on their management².

The traditional therapy for cerebral venous thrombosis is anticoagulation with heparin followed by oral anticoagulation. This helps to stop the thrombotic process and prevents thromboembolic events. Concerns over venous infarction, embolisation and persistent septic thrombophlebitis have prompted recommendations for the use of anticoagulants in patients with sigmoid sinus thrombosis². Bradley et al.²⁶ suggested that patients with thrombosis confined to the sigmoid sinus should not be given anticoagulants to avoid the associated risks. They emphasise the importance of serial imaging with MRI, MR venography, or CT venography in all patients being monitored for thrombus progression²⁰. The call for anticoagulation is supported by evidence of thrombus progression, extension to sites other than those seen on initial examination (such as the proximal internal jugular vein and the transverse or cavernous sinus), neurological changes, persistent fevers and embolic events². Endovascular thrombolysis can be attempted, administering a thrombolytic enzyme (usually urokinase). Published reports only include case reports and uncontrolled studies, from which it is impossible to say whether the results obtained with endovascular thrombolysis are superior to those achieved with systemic heparin²⁷.

Conclusions

Planned closure of the sigmoid sinus was performed without any consequences in all our 218 cases. The sinus was always sacrificed when the procedure involved the jugular foramen, as well as in cases of *en bloc* STBR. Tumours of the jugular foramen and adjoining sites may have at least partially closed the sigmoid sinus prior to any surgery, with new collateral venous pathways developing before any surgical closure is performed. To assess the safety of sigmoid sinus closure, we critically reviewed our experience and the literature on the venous drainage patterns in the brain and skull base. The vicarious role of the emissary veins and vertebral plexus was investigated, as was the mechanism behind symptomatic intracranial hypertension. Our literature review showed that there is a shortage of experimental studies in humans, but supported our findings, i.e. that planned closures of the sigmoid sinus are clinically irrelevant providing they are done after venous discharge from the skull base and brain has been reliably assessed.

References

- 1 Zanoletti E, Martini A, Emanuelli E, et al. *Lateral approaches to the skull base*. Acta Otorhinolaryngol Ital 2012;32:281-7.
- 2 Handley TP, Miah MS, Majumdar S, et al. *Collet-sicard syndrome from thrombosis of the sigmoid-jugular complex: a case report and review of the literature*. Int J Otolaryngol 2010;2010:203587.
- 3 Kanno T, Kasama A, Suzuki H. *Safety of ablation of the sigmoid and transverse sinuses: an experimental study*. Skull Base Surg 1993;3:146-51.
- 4 Mullen MT, Sansing LH, Hurst RW, et al. *Obstructive hydrocephalus from venous sinus thrombosis*. Neurocrit Care 2009;10:359-62.
- 5 Mortazavi MM, Tubbs RS, Riech S, et al. *Anatomy and pathology of the cranial emissary veins: a review with surgical implications*. Neurosurgery 2012;70:1312-9.
- 6 Mazzoni A. *The petro-occipital trans-sigmoid approach for lesions of the jugular foramen*. Skull Base 2009;19:48-56.
- 7 Mazzoni A, Danesi G, Zanoletti E. *Primary squamous cell carcinoma of the external auditory canal: surgical treatment and long term outcome*. Acta Otorhinolaryngol Ital 2014;34:129-37.
- 8 Eckley W. *Anatomic and physiologic correspondences of child and adult*. J Am Med Assoc 1903;40:1355-8.
- 9 Nathoo N, Caris EC, Wiener JA, et al. *History of the vertebral venous plexus and the significant contributions of Breshet and Batson*. Neurosurgery 2011;69:1007-14.
- 10 Ciuti G, Righi D, Forzoni L, et al. *Differences between internal jugular vein and vertebral vein flow examined in real time with the use of multigate ultrasound color doppler*. AJNR Am J Neuroradiol 2013;34:2000-4.
- 11 Batson OV. *Anatomical problems concerned in the study of cerebral blood flow*. Fed Proc 1944;3:139-144.
- 12 Kinal ME. *Hydrocephalus and the dural venous sinuses*. J Neurosurg 1962;19:195-201.
- 13 Behari S, Tyagi I, Banerji D, et al. *Postauricular, transpetrous, presigmoid approach for extensive skull base tumors in the petroclival region: the successes and the travails*. Acta Neurochir (Wien) 2010;152:1633-45.
- 14 Suzuki M. *Histopathological and enzyme-histochemical changes after brain compression*. Neurol Med Chir 1981;21:221-32.
- 15 Tsutsumi K, Shiokawa Y, Sakai T, et al. *Venous infarction following the interhemispheric approach in patients with acute subarachnoid hemorrhage*. J Neurosurg 1991;74:715-9.
- 16 Bigelow DC, Hoffer ME, Schlakman B, et al. *Angiographic assessment of the transverse sinus and vein of labbe to avoid complications in skull base surgery*. Skull Base Surg 1993;3:217-22.
- 17 Ohata K, Haque M, Morino M, et al. *Occlusion of the sigmoid sinus after surgery via the presigmoidal-transpetrosal approach*. J Neurosurg 1998;89:575-84.
- 18 Leblebisatan G, Yis U, Dogan M, et al. *Obstructive hydrocephalus resulting from cerebral venous thrombosis*. J Pediatr Neurosci 2011;6:129-30.
- 19 Mamikoglu B, Wiet RJ, Esquivel CR. *Translabyrinthine approach for the management of large and giant vestibular schwannomas*. Otol Neurotol 2002;23:224-7.
- 20 Brackmann DE, Green JD. *Translabyrinthine approach for acoustic tumor removal*. Otolaryngol Clin North Am 1992;25:311-29.
- 21 Swift GW. *The transverse sinus and its relation to choked discs*. 1930;3:47-70.
- 22 Galgano MA, Deshaies EM. *An update on the management of pseudotumor cerebri*. Clin Neurol Neurosurg 2013;115:252-9.
- 23 Ideguchi M, Kajiwara K, Yoshikawa K, et al. *Benign fibrous histiocytoma of the skull with increased intracranial pressure caused by cerebral venous sinus occlusion*. J Neurosurg 2009;111:504-8.
- 24 Sebire G, Tabarki B, Saunders DE, et al. *Cerebral venous sinus thrombosis in children: risk factors, presentation, diagnosis and outcome*. Brain 2005;128:477-89.
- 25 Spetzler RF, Dasplit CP, Pappas CT. *The combined supra- and infratentorial approach for lesions of the petrous and clival regions: experience with 46 cases*. J Neurosurg 1992;76:588-99.
- 26 Bradley DT, Hashisaki GT, Mason JC. *Otogenic sigmoid sinus thrombosis: what is the role of anticoagulation?* Laryngoscope 2002;112:1726-9.
- 27 Canhao P, Falcao F, Ferro JM. *Thrombolytics for cerebral sinus thrombosis: a systematic review*. Cerebrovasc Dis 2003;15:159-66.

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