Surgical History of Sleep Apnea in Pediatric Patients with Chiari Type 1 Malformation

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Introduction

Chiari malformation type 1 (CM-1) has become increasingly recognized as a significant clinical burden in approximately 3.6% of children undergoing brain and cervical spinal cord imaging. Sleep related breathing disruption can result from compression of the medullary respiratory control centers and manifest in central or obstructive sleep apnea, hypoventilation, or even sudden death. Without a widely recognized, definitive correlation between magnitude of tonsillar herniation and clinical manifestations of cervicomedullary junction compression, debate lingers over the indications for operative versus nonoperative management of CM-1. In order to gain insight into surgical technique and potential implications of sleep apnea in the management of CM-1, the present study assessed two different posterior fossa decompression techniques (duraplasty and dural splitting) and long-term symptomatic and radiographic outcomes for surgically managed pediatric patients with concurrent sleep apnea and CM-1.

Methods

The authors retrospectively reviewed eight consecutive cases of pediatric patients less than 18 years old with symptomatic sleep related breathing disorders and concurrent CM-1 (herniation of cerebellar tonsils at least 5 mm below foramen magnum). Patients underwent comprehensive multidisciplinary evaluation including child neurology, sleep specialist and otolaryngology consultations. Surgical technique was reviewed and correlated to long-term radiographic and clinical outcomes.

Results

Surgical intervention was associated with improvement of symptoms and radiographic evidence of tonsillar ectopia and syringomyelia. Overall, surgery improved sleep apnea in 62.5% of patients (100% of the duraplasty group and 50% of the dural splitting group) (p=0.43). Although duraplasty and dural splitting were both associated with a notable reduction in tonsillar herniation of 58% and 35%, respectively, there was no significant difference in radiographic findings over a median follow-up period of 45.7 months.

Conclusions

Duraplasty and dural splitting are associated with radiographic and symptomatic improvement in pediatric patients with concurrent sleep-related breathing disorders and CM-1. Sleep apnea represents an indication for surgical decompression in these patients.

Learning Objectives

By the conclusion of this session, participants should be able to:

1. Understand sleep apnea as a relative indication for posterior fossa decompression in pediatric patients with Chiari type 1 malformation
2. Recognize duraplasty and dural splitting offer comparable rates of radiographic and symptomatic improvement
3. Appreciate that intraoperative ultrasound can be beneficial in determining restoration of cerebrospinal fluid (CSF) circulation in the posterior fossa
4. Acknowledge longitudinal follow-up studiy of patients with neurological deficits and/or severe symptoms to further elucidate CM-1 natural history

References

Benglis et al, J Neurosurg Pediatr 7:375-379, 2011; Chauvet et al, Neurosurg Rev 32:465-470, 2009; Dauvilliers et al, J Neurol Neurosurg Psychiatry 78:1344-1348, 2007; Dhamija et al, Clin Neurol Neurosurg 115:1837-1841, 2013; Durham et al, J Neurosurg Pediatr 2:42-49, 2008; Limonadi et al, J Neurosurg 101:184-188, 2004; Munshi et al, Neurosurgery 46:1384-1389; discussion 1389-1390, 2000; Novegno et al, J Neurosurg Pediatr 2:179-187, 2008; Yeh et al, J Neurosurg 105:26-32, 2006

Figure 1. Duraplasty

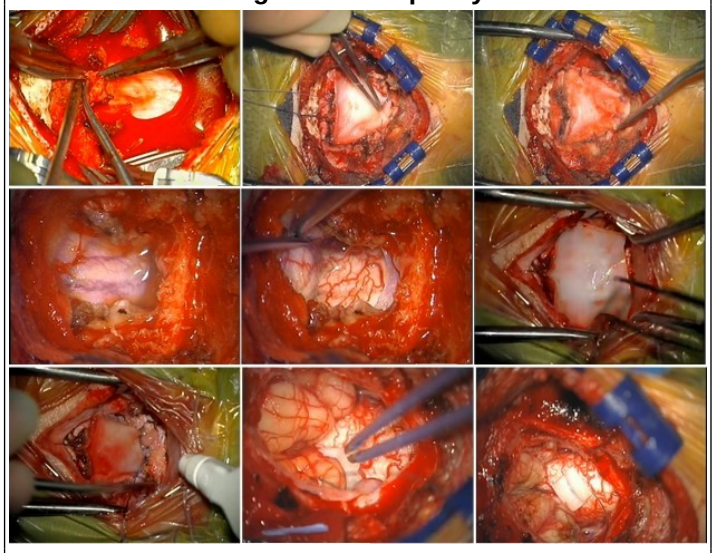

Fig 1: removal of thick fibrous dural band. Fig 2-3: dural tack-up suture. Fig 4-5: open arachnoid, no adhesions. Fig 6-7: duraplasty. Fig 8-9: tonsillopexy

Figure 2. Dural splitting

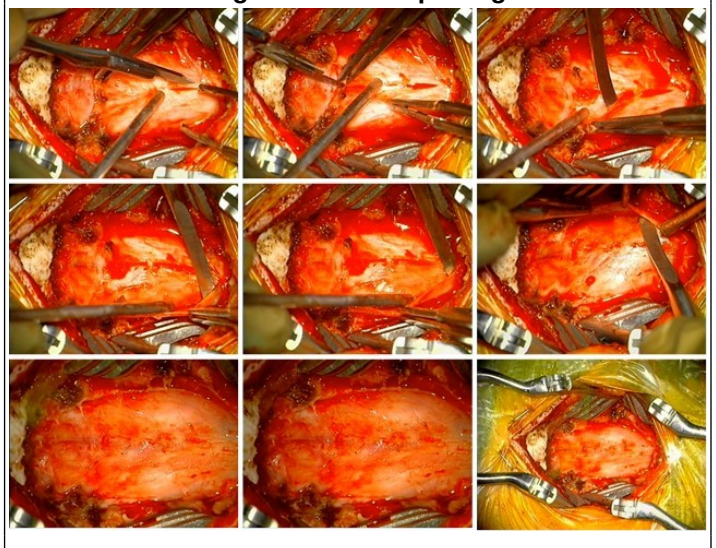

Fig 1: incising dura. Fig 2: splitting dural band. Fig 3-6: blunt followed by sharp dissection. Fig 7-8: thinned dura. Fig 9: translucent dura