Stability-Sparing Endoscopic Endonasal Odontoidectomy in a Malformative Craniovertebral Junction: Case Report and Biomechanical Considerations.


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Abstract

BACKGROUND:
The craniovertebral junction (CVJ) is often involved in a wide range of congenital, developmental and acquired pathologies that can create bony and ligamentous instability or cause direct compression on the medulla and cervical spine cord, resulting in significant impairment. Atlas assimilation is the most common malformation in the CVJ and can be frequently associated with basilar invagination (BI) and Chiari malformation (CM) type I. Posterior atlas assimilation more frequently leads to BI type II with a mass effect on neural structures but usually no signs of biomechanical instability. Operative approaches to the CVJ have undergone a remarkable evolution and can be divided into ventral, lateral and dorsal ones. In this kind of surgery, it is vital to detect and eventually treat any CVJ instability.

CASE DESCRIPTION:
We present a case of CVJ malformation comprising assimilation of the posterior arch of the atlas, BI type II and CM, treated by endoscopic endonasal odontoidectomy and partial clivus removal to spare CVJ stability.

CONCLUSION:
Neurological and biomechanical analysis of all CVJ malformations permits stratification and selection of those cases that can be managed by simple, direct, minimally invasive decompression with no need for surgical fusion.

KEYWORDS:
Atlas assimilation; Basilar invagination; Chiari I malformation; Cranio-cervical malformation; Cranio-vertebral junction; Endoscopic endonasal odontoidectomy

INTRODUCTION:
The association of Hirschsprung disease (HSCR) and Down Syndrome (DS) is not uncommon (HSCR+DS). This paper aims at reporting the results of a 24-year series focusing on surgical approach, complications and long term outcome.

MATERIALS AND METHODS:
The notes of all patients admitted with a diagnosis of HSCR+DS have been retrospectively reviewed. Surgical details, intraoperative complications, long term issues and functional outcome have been recorded. The results have been compared to those of patients without DS and were assessed based on surgical approach.

RESULTS:
A total of 23 HSCR+DS out of a series of 385 HSCR (6%) have been included. Preoperative enterocolitis (HAEC) was reported by 32%. Associated anomalies were detected in more than half of the patients. In particular, Congenital Heart Defects (CHDs) were reported by 57%. Postoperative complications (mostly symptomatic anal sphincter achalasia) were experienced by 55%. Constipation was experienced by 30%; severe continence issues, by 53%. One patient suffering from severe CHDs died. With regard to complications, only symptomatic anal achalasia requiring intrasphincteric BoTox injection was significantly more frequent in HSCR+DS (30% vs 10%, p = 0.0071). Similarly, continence proved to be significantly worse in HSCR+DS.

DISCUSSION:
With the exception of symptomatic anal achalasia, HSCR+DS patients proved not to have a higher likelihood of complications compared to HSCR alone. On the other hand, functional results in the long term are worse. As a consequence, long term follow up and personalized rehabilitation programs are warranted for this delicate subset of HSCR patients.

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Continence; Down; Enterocolitis; Hirschsprung; Minimally invasive surgery