Diastematomyelia: Our Experience at Rady's Children's Hospital

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Introduction

Diastematomyelia is a rare congenital occurrence defined as a sagittal division of the spinal cord into two hemi cords with an incidence of 300,000 newborns per year. It is a consequence of an abnormal adhesion between ectoderm and endoderm with persistent accessory neurenteric canal between the yolk sac and amnion, reflecting the failure of the neural tube to close.

Diastems can manifest themselves where the hemi cords are in two separate dural sheaths separated by either an osseocartilaginous median septum or a fibrous septum. In 60% of patients with diastematomyelia, the two hemicords are each covered by a single dural sac.

The progressive dysfunction may lead to a combination of cutaneous, orthopedic, urologic, and neurologic signs and symptoms. Surgery can only address mass effect and, in most cases cord, tethering, but it does not address congenital myelodysplasia or predictably reverse established clinical defects.

Methods

We performed a retrospective analysis of 30 patients presenting

Results

Among the 30 patients there were 20 females and 10 males (mean age = 6.1 years). 28 patients had confirmed association with tethered cord and Lipomatous malformations were confirmed in 12 patients. On follow-up, 23 patients underwent surgical intervention with no complications and remained neurologically at baseline. Five patients Improved, 1 had a transient decline in motor function, and 1 had a decline in sensory function following surgery.

Conclusions

The presentation spectrum of dysatematomyelia can be complex ranging from progressive scoliosis or neurologic decline as opposed to being asymptomatic. Early clinical recognition, radiographic diagnosis, and neurosurgical treatment are essential to prevent complications and stop progression. Neurologic compromise following surgery is not common despite the complexity of the cases, and when present does not result in impaired autonomy. Thus, the benefit risk ratio indicates the need for the consideration of potential surgical intervention upon diagnosis.

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe types of Diastematomyelia 2) evaluate the benefit risk ratio indicates the need for the consideration of potential surgical intervention in Diastematomyelia cases

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