Original articles

Cricopharyngeal incoordination in infancy

Stojan M. Živković
Hospital for Pediatric Surgery, Belgrade University Hospital, Tiršova 10, Y-11000 Belgrade, Yugoslavia

Abstract. Cricopharyngeal incoordination in newborns and infants is a polyetiological condition. From 1973 to 1986, 13 patients with defective swallowing mechanisms were seen at our hospital. The patients were divided into two groups. The first group, consisting of 8 patients with transient cricopharyngeal incoordination due to immaturity and birth trauma, did not require surgical intervention. The second group, composed of 5 patients with permanent incoordination, required surgical treatment. Cricopharyngeal achalasia and familial autonomic dysfunction (Riley-Day syndrome) were the most frequent causes in this group. We thought it wise to simplify feedings by using a gastrostomy, which we also used for retrograde dilatation. Permanent cricopharyngeal incoordination may be treated by dilatation or myotomy. Familial dysautonomia required additional surgical procedures, depending on the associated upper gastrointestinal disorder. Indications for myotomy of the cricopharyngeal muscle in infancy have not been clarified to date. Dilatation may improve the passage of food through the upper esophagus, but if this procedure fails, myotomy is indicated.

Key words: Cricopharyngeal incoordination, transient and permanent — Achalasia — Familial dysautonomia

Introduction

Over the last 2 decades, several authors have reported cases of cricopharyngeal incoordination in the pediatric age group [3, 5, 10]. This entity is well known in adults, but has occasionally been described among newborn infants [12, 13]. The condition should be suspected in the newborn with problems of choking, coughing, and cyanosis. Cineradiographic examination is essential for diagnosis and to exclude anatomical abnormalities such as a congenital H-type tracheoesophageal fistula, a vascular ring, congenital esophageal stenosis, deficient posterior laryngeal wall, etc. [6]. Brain damage and neurological deficits must be also excluded.

Cricopharyngeal incoordination of the newborn may occur either transiently or permanently. The former disappears spontaneously after maturation of the reflex within the first 2 weeks of life [3, 5]. By the end of this period the newborn develops the ability to swallow a bolus normally, without tracheal or bronchial aspiration. However, patients with cricopharyngeal incoordination of prolonged duration require different types of surgical management [4, 7, 9]. Achalasia and familial dysautonomia were found most frequently in this group. Familial dysautonomia has special needs, and requires additional procedures including gastrostomy, Nissen's fundoplication, and pylorotomy. These procedures cannot cure the dysautonomic disease, but they can reduce recurrences of pneumonia [1, 8, 11].

Materials and methods

During the period from 1973 to 1986, 13 patients with dysphagia were seen at our hospital. Ten of the 13 patients developed clinical findings within the first 3 days of delivery. In all patients saliva accumulated in the mouth, causing cyanosis. Respiratory distress and aspiration pneumonia were present in 4 patients; regurgitation through the nose was noted in 3. Cineradiographic examinations were performed twice in the early newborn period, the first for excluding rare anatomical abnor-
Fig. 1 A, B. Case 1. A Lateral esophagogram with spasm of the cricopharyngeal muscle (Lower arrow). The upper arrow shows contrast material in the nasal cavity. B Fifteen days later spasm no longer present.

Fig. 2. Case 13. A Lateral esophagogram: upper arrow = typical spasm; lower arrow = trancheal and bronchial aspiration. B, C Cineradiographic finding 5 years after left myotomy shows good barium passage, but slight spasm is present (arrow).