Achalasia in Pediatric Population: Use of High-Resolution Manometry in Children, Achalasia in Pediatric Population

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Abstract

Achalasia is a rare esophageal motor disorder in the pediatric population, in pa

Figure 1: High-resolution esophageal pressure topography (EPT) study in a 11-year- old boy (case 1) with weight loss and dysphagia. The plot shows pan-esophageal pressurization and impaired

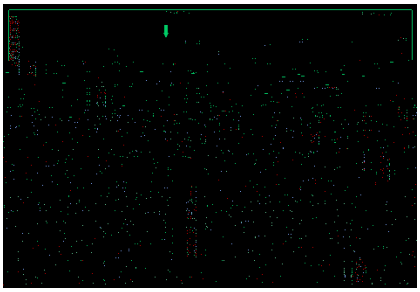

Figure 4: Conventional manometry in a 7-year-old girl showing hypertensive lower esophageal sphincter (LES), mean pressure of 50 mmHg, with incomplete relaxation of the LES (residual LES pressure of 9 mmHg) during wet swallow (arrow), aperistalsis in the body of the esophagus (observed low amplitude waves and aberrant morphology mirror image).

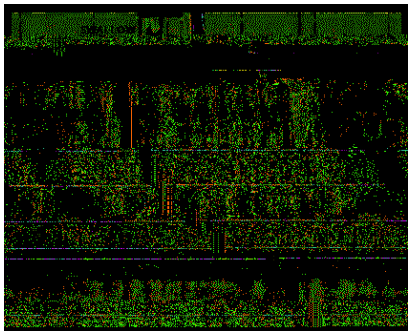

Figure 5: High-resolution esophageal pressure topography (EPT) showing a 17-years girl (case 3), after 10-year Heller myotomy. EPT with normal lower esophageal sphincter pressure and complete relaxation, aperistalsis esophageal body and normal pharyngo-esophageal coordination.

Discussion

Achalasia is a rare neurodegenerative esophageal disorder in pediatric population. The disease is rarer in children under 5 years of age. The incidence in childhood is 0.11 / 100,000 children per year. Overall, fewer than 5% of patients with symptoms are under 15 years. The disease is more prevalent in males and is often idiopathic. Achalasia has been associated with trisomy 21, congenital hypoventilation syndrome, glucocorticoid insufficiency, eosinophilic esophagitis, familial dysautonomia, Chagas disease, and "AAA" syndrome (Achalasia, alacrima and insensitivity to ACTH), [1,3,8,9].

The pathophysiology is characterized by the selective loss of inhibitory neurons located in the myenteric plexus, resulting in alterations in relaxation of the lower esophageal sphincter (LES) and absence of peristalsis in the esophageal body. Loss or absence of myenteric ganglion cells, destruction or chronic inflammation, support the theory that abnormalities in the parasympathetic innervation of the esophagus result in disturbances of esophageal motility; however, the precise etiology of this disorder is still poorly understood [10]. In pediatric population, symptomatology is diverse, often having progressive dysphagia, which is the main reason for medical consultation, vomiting and weight loss [7], as reported in this series of cases. Young children and infants may present atypically as recurrent pneumonia, nocturnal cough, aspiration events, hoarseness and difficulty feeding; it is often misdiagnosed as GERD. Children may have delayed development, eating disorders or asthma, which in turn leads to a delay in diagnosis for a period of 6-10 years. Up to 50% of children are treated with antiacids or prokinetic before making the diagnosis of achalasia [9].

The diagnosis is based on clinical data, using the esophagogram (which typically shows a dilated esophagus with image of "bird´s beak" in distal esophagus), and confirmed with esophageal manometry [2,5,7]. Esophageal manometry is the gold standard in diagnosing achalasia, the high-resolution manometry (HRM) is leading to the