DIAGNOSIS AND MANAGEMENT OF ESOPHAGEAL ACHALASIA

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INTRODUCTION

- **Esophageal achalasia** is a rare **neurodegenerative motility disorder** characterized by spasm or failure to relax of the lower esophageal sphincter (LES).
- Clinical presentation includes **dysphagia** (>90%) for solid and liquids, regurgitation of undigested food, nocturnal cough, chest pain, and weight loss.
- Diagnosis is typically delayed 2-3 years from symptoms onset, due to frequent confusion with more common entities such as gastroesophageal reflux disease. It is based on **clinical presentation, esophageal manometry, barium esophagogram, upper gastrointestinal tract endoscopy**.
- **Endoscopic and surgical treatment options** remain the main therapeutic options.

Fig 1. Barium swallow demonstrating esophageal dilatation and narrowing at the LES ('bird like' deformity).

Fig 2. Esophageal manometry. **A.** Increased basal lower esophageal sphincter pressure. **B.** Simultaneous esophageal body contractions-absent peristalsis. **C.** Incomplete LES relaxation.

RESULTS

- 3 children with achalasia were diagnosed in University General Hospital Attikon, including 2 males and 1 female with an average age of 8 years (range: 5-14 years).
- The **duration of the disease** before the diagnosis was about 2 years.
- The **diagnosis** of achalasia based on clinical symptoms of one or more of the following presentations: vomiting/regurgitation, dysphagia for solids and liquids, retrosternal pain, and weight loss.
- **Barium meal examination** showed diagnostic signs of achalasia: an esophageal dilation and a 'bird-beak' at the cardia (Figure 1).
- **Esophago-gastroduodenoscopy** demonstrated dilated esophagus in all of them and esophagitis in 1 patient.
- **Conventional esophageal manometry** revealed incomplete relaxation of LES and abnormal esophageal peristalsis (Figure 2).
- **High resolution manometry** (HRM) showed achalasia type II in two cases and type III in the third patient (Chicago classification).
- Patients were treated **surgically** to reduce the pressure of LES. All of them received Heller's esophagomyotomy with antireflux procedure, while one of them required additional pneumatic dilatation.
- The mean follow-up time was 24 months. The outcome was perfect in 2 of them without any symptoms while the last one dropped out of follow-up.

CONCLUSIONS

- Diagnosis of esophageal achalasia requires thorough history and dedicated esophageal testing.
- HRM is able to classify achalasia and predict disease outcome.

References

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