

**ESPGHAN:** *European Society for Paediatric Gastroenterology, Hepatology and Nutrition.*

**NASPGHAN:** *North American Society for Paediatric Gastroenterology, Hepatology and Nutrition.*

*Both are multi-professional organisations whose aim is to promote best practice in the delivery of care for the health of children and adolescents with special attention to the gastrointestinal tract, liver and nutritional status.*

## **ESPGHAN-NASPGHAN Guidelines for the Evaluation and Treatment of Gastrointestinal and Nutritional Complications in Children with OA/TOF**

*A brief look by Sue Lewis-Jones*

### **Adult TOFs keep reading!**

While the title of these new guidelines refers to “children”, the content does go on to include advice and information for adolescents transitioning from paediatrics into adult services and for Adult TOFs.

These are an exciting development. For the first time, there are guidelines and advice written for medics and health professionals, by medics and health professionals, regarding the treatment and care of people born with the TOF condition.

They acknowledge that post-operative outcomes have changed since the early days of OA/TOF repair in the 1940s and that with improved techniques and care, the focus has shifted from survival to looking at future quality of life issues. “OA is no more just a neonatal surgical problem but a lifelong problem.”

They list the problems most frequently faced by those born with the TOF condition, as not only prevalent in childhood but also in adulthood: respiratory problems, nutritional and gastrointestinal issues such as gastro-oesophageal reflux, anastomotic strictures, feeding disorders, dysphagia, oesophageal dysmotility, peptic oesophagitis, gastric metaplasia and Barrett’s oesophagus.

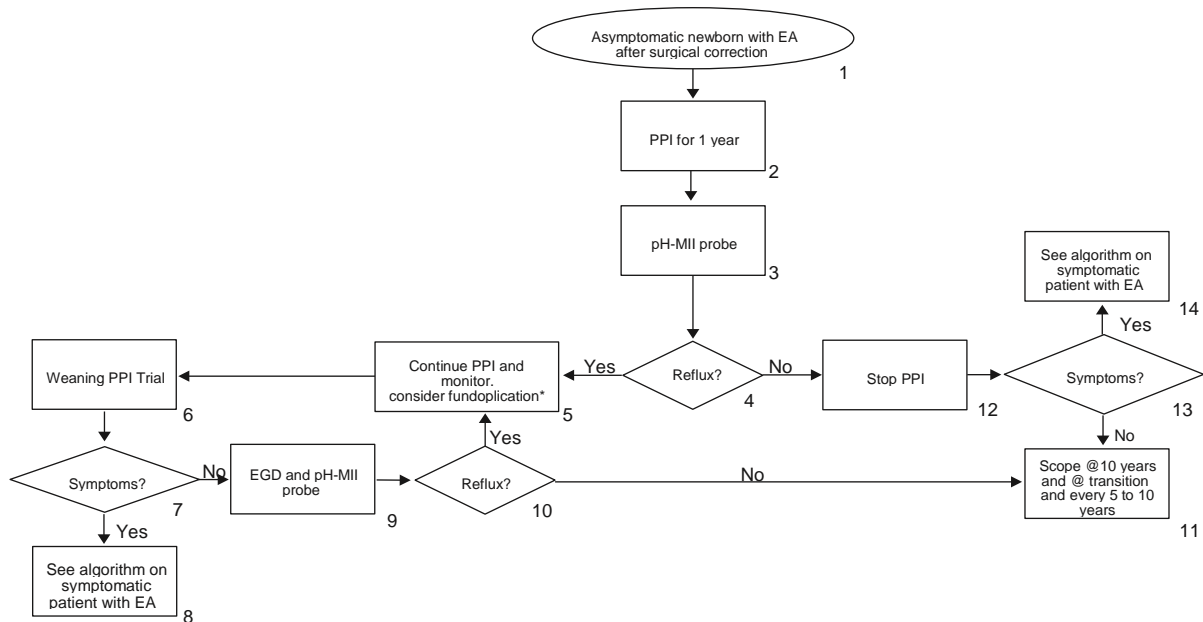
Interestingly, they say that while it was acknowledged (perhaps amongst specialists) that some TOF patients faced greater difficulties with these and that there was a need for a multi-disciplinary follow-up, until recent years there have been no recommendations available on care.

The International Network on Esophageal Atresia (INoEA) formed in 2013 to help formulate these guidelines. (INoEA organises medical conferences on OA/TOF, the last held in 2016 in Sydney, Australia.) A working party of paediatric surgeons and gastroenterologists started looking at key areas: reflux, its treatment, testing and surgery; dysmotility; cyanotic spells; dysphagia; anastomotic stricture; eosinophilic oesophagitis; transition to adulthood; long-term digestive issues; surveillance; and querying whether quality of life was in some cases impaired.

### **On reflux**

Many babies experience reflux but in the TOF population it can persist long-term and can cause many problems, affecting not only the oesophagus but also the airways, and causing failure to thrive. The report suggests that reflux is still a problem in TOF children after the age of two, even in those who previously did not have symptoms. It can persist throughout life, making the likelihood of oesophagitis high throughout childhood and adulthood. One of the long-term complications associated with untreated reflux is Barrett’s oesophagus.

This can affect quality of life and one of the guidelines suggests regular monitoring to check for the presence of acid reflux. It recommends that monitoring should take place before any decision to stop anti-acid medications, even when there appear to be no symptoms and also after a fundoplication, as reflux can still reoccur. It also recommends that eosinophilic oesophagitis (EoE) should be ruled out before a fundoplication is considered.



The ESPGHAN/NASPGHAN flow chart shows recommended pathways for treating reflux.

- Reflux should be treated to prevent complications and strictures up to the age of one year and then longer where it persists.
- Regardless of symptoms, there should be one scope after stopping PPI medication, one before the age of ten years, another at transition and thereafter, every five to ten years.

### On dysphagia

The report looked at dysphagia, which are swallowing problems, and how the oesophagus functions in people born with OA. They estimated that dysphagia is far more widespread in the TOF population than reported. Children in particular, never having known anything different, do not regard their problems as abnormal and have learnt to adapt. The report suggests that dysphagia continues to be experienced indefinitely, no matter how long after repair surgery.

### On transition and adulthood

Cameron Haight carried out the first successful repair in 1941 and so people born with OA are for the first time approaching their seventies. The report recommended that focus on the long-term outcomes of these patients is necessary.

Most TOFs leave paediatric services with no follow-up or continuity of care. Yet studies have shown that gastro-intestinal problems (and, to a lesser degree, respiratory difficulties) are common amongst them, affecting a much higher proportion than in the ordinary population.

Swallowing problems are frequent and affect 39%–85% of Adult TOFs compared to around 2% in a control group.<sup>1</sup> Reflux also affects a much higher proportion of Adult TOFs than in the general population. Taylor et al found that reflux symptoms were reported in 63% of TOF subjects with a quarter of those having severe reflux symptoms (determined as happening on at least three days a week).<sup>2</sup>

Adult TOFs will have become very used to what is “normal” for them, living with the symptoms of reflux and dysphagia for so long that they often do not seek medical advice, are therefore under-medicated and, consequently, their symptoms worsen. Studies have indicated that as few as 10% of Adult TOFs take proper medication for their reflux. Where reflux remains untreated, it leads to oesophagitis and further problems such as Barrett’s oesophagus. There is a higher incidence of oesophagitis and Barrett’s oesophagus in adults with TOF/OA compared to the general population.

Current studies show no increase in oesophageal cancer in Adult TOFs but it does remain a concern.

- The guidelines recommend that there should be transition from paediatric to adult services with care passing to a physician with knowledge of the complexities of TOF/OA.
- Thereafter, they recommend regular clinical follow up and routine endoscopy with biopsies every five to ten years for every Adult TOF (especially those with symptoms of dysphagia, reflux, respiratory problems or anaemia.)
- There should be additional endoscopy on the development of new or worsening symptoms.

***For further information do come along to the next TOFS Conference (Luton, Saturday 14 Oct 2017) when Professor Gottrand, President of INoEA, will be speaking about these guidelines. Look at the full report on the ESPGHAN/NASPGHAN websites.***

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<sup>1</sup> Sistonen SJ, Koivusalo A, Nieminen U, et al. Esophageal morbidity and function in adults with repaired esophageal atresia with tracheoesophageal fistula: a population- based long-term follow-up. *Ann Surg* 2010;251:1167–73.

<sup>2</sup> Taylor AC, Breen KJ, Auldish A, et al. Gastroesophageal reflux and related pathology in adults who were born with esophageal atresia: a long-term follow-up study. *Clin Gastroenterol Hepatol* 2007;5:702–6.