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OESOPHAGEAL ATRESIA AND WEANING

Kathryn Lowes
Paediatric Dietitian
Great Ormond Street Hospital for Children

Oesophageal atresia (OA) occurs in one in 2,500 to 4,500 births. It is a congenital abnormality whereby the oesophagus ends blindly in a pouch, resulting in a non-continuous route from the mouth to the stomach. It occurs in isolation in only seven percent of infants; however, in 86 percent of cases it is diagnosed with a distal trachea-oesophageal fistula (TOF) also present (1).

In 50 percent of cases, OA +/- TOF will present with anomalies such as VACTERL association (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies and limb abnormalities), CHARGE syndrome (Coloboma of the eye, heart defects, atresia of the choanae, retardation of growth and/or development, genital and/or urinary abnormalities and ear abnormalities and deafness), Trisomy 13, DiGeorge syndrome, along with others.

PRESENTATION AND DIAGNOSIS

OA can be suspected on an antenatal scan, but a confirmed diagnosis will be made after birth. An infant with OA +/- TOF will classically present with respiratory distress, choking, feeding difficulties and frothing at the mouth in the first few hours after birth.

PRE-SURGERY MANAGEMENT AND SURGERY

Essentially the management of an OA/TOF involves surgical correction of the anomaly. Until surgery, management is to provide nutritional support and hydration and to prevent aspiration. A multidisciplinary team approach is required throughout the infant's journey; from pre-repair to the teenage years, as many children will go on to have further complications and interventions. This should involve, surgeons, nursing staff, physiotherapists, dietitians and speech therapists (2, 3).

Surgery to repair the anomaly will take place as soon as possible, but the distance between the proximal and distal ends of oesophagus needs to be short enough to form a primary anastomosis. The surgeons will often pass a trans-anastomotic tube (TAT), allowing a continuous route for milk through to the stomach and for gastric decompression.

In some infants, the distance between the two ends is too great; this is called 'long gap oesophageal atresia'. The way this is managed differs between hospitals; in some cases a temporary cervical oesophagostomy is formed to allow the infant to swallow saliva and a gastrostomy is placed for feeding. Children can wait up to six months for a repair. Often 'sham feeding' is recommended to prevent oral aversion while the infant waits for the surgical repair. Sham feeding involves a 'Replogle Tube' connected to suction to drain the milk from the upper oesophageal pouch, thus preventing aspiration. Sham feeding is used to promote the establishment of oral feeding either by breast or bottle, prior to repair of the OA (1).

Kathy has been working at Great Ormond Street for the last three years. She has spent the last 18 months working in general surgery and intestinal failure, which has involved working with children with complex feeding difficulties.

Infants and toddlers will not chew their food to begin with, so it is best to avoid foods that could get stuck until chewing and oral development have improved.

If oral feeding is delayed for more than even just a few weeks, oral sensitivity and vomiting can become a problem and this can take a long time to resolve. So where there is a delay in repairing the OA and if an oesophagostomy has been placed, it is good practice to start sham feeding to prevent oral aversion.

Infants usually grow well on breast milk or standard infant formula if they are able to receive sufficient volumes.

POST-SURGERY AND WEANING ONTO FOODS

Once the oesophagus has been repaired, there are ongoing problems that will occur in most children, though some children will not have any problems. Below are some of the complications that can occur post-surgery.

- Stricturing - narrowing at the anastomosis, often requiring a dilatation procedure.
- Oesophageal dysmotility - can continue into adult life.
- Oesophageal obstruction - food is not propelled down to the stomach, due to an incoordination of the oesophageal muscle or due to a stricture and a mechanical obstruction. If the blockage does not clear with sips of water, this will require admission to the local hospital to have the piece of food removed. This is likely to become less common as the child gets older.
- Gastro-oesophageal reflux (GOR) - if a child is refusing food regularly, further investigations into this will be necessary, even if they are not vomiting (2, 4).

Evidence suggests that oesophageal foreign body obstruction does occur in a number of patients who have had OA/TOF and parents should be cautious of lumpy, sticky foods up until five years of age and especially when different food textures are being introduced (this will depend on the individual as some patients may be on a puree/soft diet and/or even an enteral feed for longer) (5).

Weaning is an essential milestone in any child's life and, where possible, this should not be delayed. It is important for dietitians and often speech therapists to work closely with the child and to liaise with the surgical team about what foods the child can have safely. The introduction of solid foods may be particularly useful if the child has GOR, as food is heavier/denser than liquid and less likely to cause reflux.

Advice for children post-OA/TOF repair will depend very much on the individual. Where possible, the infant should start weaning at the usual age of around six months on stage 1 foods. Emphasis should be put on taking it slowly and going at the baby's pace, as moving on to a greater variety of textures will be a longer, slower, process in children who have had OA +/-TOF (2). The role of the dietitian is to ensure that the diet is nutritionally complete and the child is growing on the limited textures that he/she is managing. Advice about what textures are allowed and not allowed should be given. It is important to remember every child is different and some may progress faster than others.

Infants and toddlers will not chew their food to begin with, so it is best to avoid foods that could get stuck until chewing and oral development have improved. Difficult to chew lumps, such as meats or raw vegetables and sticky/doughy foods should be avoided until three to five years of life - this will depend on the surgical team's advice and the child's progression post-surgery.

FOODS TO AVOID AND FEEDING ADVICE

There is limited research on weaning these children and so advice is largely based on the experience of parents and healthcare professionals, who have worked closely with this patient group (2, 4).

On initial introduction, adding liquid to foods and keeping a smooth consistency is advisable (a blender may be needed and a sieve used to remove the lumps). It is best to start with a thin puree and

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1 Contains 1.89g/100kcal of protein, including α-lactalbumin, making the protein level and quality closer to that found in breastmilk (1.7g/100kcal). Nommsen LA et al. Am J Clin Nutr 1991; 53: 457–465.
2 Koletzko B et al. Am J Clin Nutr 2009; 89(5):1502S–8S.
3 Price per 100g of infant milk powder: HiPP £1.06, Aptamil £1.11. Price per case of 24 infant milk hospital formula: HiPP £8.36, Aptamil £8.84. Prices correct as at April 2015.

Important Notice: Breastfeeding is best for babies. Breastmilk provides babies with the best source of nourishment. Infant formula milks and follow on milks are intended to be used when babies cannot be breastfed. The decision to discontinue breastfeeding may be difficult to reverse and the introduction of partial bottle feeding may reduce breastmilk supply. The financial benefits of breastfeeding should be considered before bottle feeding is initiated. Failure to follow preparation instructions carefully may be harmful to a baby's health. Infant formula and follow on milks should be used only on the advice of a healthcare professional.

As children get older, support is still required and children/teenagers often benefit from eating small, more regular meals and taking their time with each meal.

assess how the infant manages before thickening. A thicker consistency should be gradually given, offering a couple of spoonful's at a time to assess how the child copes. The child should be in an upright position, not force fed and moving on at the child's own pace (2, 4).

When ready to try finger foods, 'bite and dissolve' foods are a good option for introducing the child to solid foods and help develop chewing. The speech therapist should be consulted on what they would deem appropriate. This will often involve foods such as puffy crisps, Organix crisps, Wotsits, Quavers, meringues or sponge fingers.

PROBLEMATIC FOODS TO AVOID UNTIL THREE TO FIVE YEARS OF AGE

- Doughy/sticky foods - breads, doughnuts, croissants, pizza
- Lumps of meat - these are fine to give if pureed (will often be needed as a source of protein)
- Raw apple
- Raw vegetables
- Orange slices

FOODS TO TREAT WITH CAUTION

- Slippery foods - hard to control foods, such as peaches, banana slices, grapes
- Rusks - they may soften easily in the mouth, but still contain lumps when swallowed
- Fibrous foods - cooked vegetables, some whole wheat/bran cereals

As the child gets older (more than > three years of age) and the introduction of more lumpy foods is appropriate, again advice would be to take it slowly. Most children will continue to do well with foods accompanied with sauces/gravies to provide more moisture to help with their swallow.

As they move on to meats, softer types such as chicken or sausage should be given rather than beef and pork (these are more fibrous). When introducing meats, to begin with they must be minced or chopped very finely and mixed with a sauce (3).

DRINKING

Some children will find it hard to co-ordinate their swallow and especially those who had long gap OA. They will not necessarily have developed a good suck reflex; they may find it easier to use a beaker or straw for fluids (3). If a child is having difficulty with drinking, he/she may need a swallow assessment from a speech and language therapist.

As children get older, support is still required and children/teenagers often benefit from eating small, more regular meals and taking their time with each meal. Growth should be carefully monitored and the diet supplemented where needed.

SUMMARY

Every infant/child should be treated as an individual when it comes to the management of post-OA/TOF repair. A supportive multidisciplinary team is important in the management of nutrition and further oral development.

There are also charities, such as TOFs (see website below), to support these families and allow them to talk to each other. Each step should be taken slowly, with an awareness of risks and their management, while working to support the family in helping the child lead as normal a life as possible.

1 Shaw V 4th Edition Clinical Paediatric Dietetics, 2014, Chapter 8 Surgery in the Gastrointestinal Tract, pages 143-150.
2 Puntis JWL, Ritson DG, Holden CE and Buick RG. Growth and feeding problems after repair of oesophageal atresia. Archives Disease in Childhood January 1990: (65): 84-88.
3 Cimador M, Carata M, Di Pace MR, Natale G, Castiglione A, Sergio M, Corsello G, De Grazia E. Primary repair in esophageal atresia. The results of long term follow-up. Minerva Pediatrica February 2006: 58 (1): 9-13.
4 TOFs- http://www.tofs.org.uk - UK registered charity to support children with TOF/OA
5 Zigman A and Yazbeck S. Esophageal foreign body obstruction after eosphageal atresia repair. Journal of Paediatric Surgery May 2002: 37 (5): 776-778.

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Questions relating to: *Oesophageal atresia and weaning*
Type your answers below and then **print for your records** or print and complete answers by hand.

Q.1	Explain the aetiology and prevalence of oesophageal atresia (OA).
A	
Q.2	What are the anomalies associated with 50% of the OA cases (with or without tracea-oesophageal fistula)?
A	
Q.3	How does OA present in infants?
A	
Q.4	Explain the multidisciplinary approach to the management of OA before surgery.
A	
Q.5	What is sham feeding and when should it be put into practice?
A	
Q.6	Describe at least three of the complications that can occur post-surgery.
A	
Q.7	What role does the dietitian play during weaning of a child with post OA?
A	
Q.8	Which foods should be treated with caution from the weaning stage and beyond?Describe the dietetic support that is required as the child gets older.
A	

Please type additional notes here . . .