Management of Oesophageal Atresia

Background:
Oesophageal atresia is a spectrum of anomalies comprising an interruption of the continuity of the oesophagus combined with or without a persistent communication with the trachea resulting in one of the following:
- **Type A**: isolated oesophageal atresia (10%)
- **Type B**: oesophageal atresia with proximal/upper pouch tracheoesophageal fistula (<1%)
- **Type C**: oesophageal atresia and distal/lower pouch tracheoesophageal fistula (most common type) (85%)
- **Type D**: a combination of Type B and C (<1%)
- **Type E**: H Type trachea-oesophageal fistula without oesophageal atresia (4%)

The estimated incidence is 1 in 3500 births. In more than 50% of babies, oesophageal atresia is present in conjunction with other anomalies. Associated anomalies are more likely if there is isolated oesophageal atresia (type A). Vertebral, cardiac, gastrointestinal tract, genitourinary and limb malformations may be associated with this condition. If oesophageal atresia (OA) or tracheoesophageal fistula (TOF) is diagnosed, examine the neonate carefully to exclude further anomalies (such as VACTERL, CHARGE association).

If untreated oesophageal atresia leads to death secondary to malnutrition or aspiration pneumonia.

**Signs and Symptoms:**
Infants can present with the following signs or symptoms:
- Maternal polyhydramnios
- Copious oral secretions post delivery
- Inability to swallow feeds
- Coughing and choking with feeds
- Aspiration of feeds/secretions
- Inability to pass an oral or naso-gastric tube (NGT)

**Initial Management:**
Gently attempt to pass a size 8 – 10F NGT. If resistance is met, perform a chest X-ray (CXR). If the CXR suggests an OA, insert a Replogle* tube to prevent aspiration of secretions. Insert the Replogle tube orally until resistance is felt then withdraw slightly and secure. Note the length inserted and record in the patient notes. Nurse nil by mouth and with the head of the bed elevated.

*A Replogle tube is a double lumen radio opaque tube, which is used for continuous suction and irrigation of the blind upper oesophageal pouch.

**Preoperative Care and precautions:**

1. Nurse in a head-up position to minimise the risk of aspiration pneumonia and reflux of acid from the stomach through the fistula.
2. Place the Replogle tube on continuous low-pressure suction using the closed disposable suction system. To maintain patency of the Replogle tube, flush with 0.5mls of normal saline every hour. Ensure the saline is aspirated back after each flush.
3. CPAP and bag and mask ventilation should be avoided as positive pressure ventilation can cause gastric distension via the distal TOF and result in perforation. During the preoperative period, if concerned regarding
respiratory distress, caution should be taken before proceeding with endotracheal intubation. If intubation unavoidable, use the least possible ventilator pressure and alert the surgeons and neonatal consultant.

4. Take great care when examining the baby for associated cardiac anomalies. Arrange an echocardiogram preoperatively to exclude any significant congenital heart disease as well as to identify the position of the aortic arch and guide the operative approach. The surgical team should be made aware of any abnormal ECHO results.

Surgical Management
Oesophageal atresia is not strictly a surgical emergency. Timing of Surgery can be postponed until the patient is stable. The baby should be carefully assessed for the presence of any associated malformations, as this will determine the urgency of surgery. Occasionally, there are situations in which surgery is undertaken as an emergency: eg premature babies with severe RDS with OA requiring high-pressure mechanical ventilation; or when OA is associated with GI malformations, for instance; duodenal atresia or anorectal atresia. The operation of choice consists of division or ligation of the fistula (if present), followed by primary anastomosis of the oesophagus in order to restore intestinal continuity and allow normal swallowing.

In isolated oesophageal atresia (Type A), the surgical options are different: either an initial gastrostomy followed by delayed primary anastomosis or oesophageal replacement, or oesophagostomy, gastrostomy followed by delayed thoracotomy/oesophageal replacement.

Postoperative care:

1. Postoperatively, there will be a trantasenomotic tube (TAT) in place if primary anastomosis of the oesophagus has been performed. The TAT acts as a stent for the repair site, as well as for nasogastric feeding once commenced.
2. Ensure that the TAT is well secured and identified and it should not be removed inadvertently. If the TAT is accidentally removed, do not reinsert, inform the surgeons. Reinsertion may cause trauma to anastomotic site.
3. Gastric aspirates should be measured, described, and recorded as for any nasogastric tube.
4. If the gap between the oesophageal elements is large, the oesophageal repair may be under tension. Nurse in a supine position with a flexed head to help reduce tension on the anastomotic site. These neonates will often require sedation and paralysis to prevent undue stretching of the anastomotic site. Post op there may be a chest drain in situ. This is usually an extra pleural drain and not placed under suction unless specifically requested by the surgical team. Even low-pressure suction may cause damage to the newly repaired oesophagus.
5. TAT feeds may be commenced as early as 24-48 hours post op unless contraindicated by the surgical team, A contrast study may be performed approximately at 5-7 days postoperatively to detect any anastomotic leak. If the contrast study is normal, oral feeds may be introduced.
6. Keep the head of the bed elevated when feeds are commenced. A degree of oesophageal dysmotility and associated gastro oesophageal reflux is commonly seen in the postoperative period. Elevating the head of the bed and utilising positioning techniques may help to control reflux symptoms. However, most patients also require antireflux treatment: Ranitidine.

Outcome:
Although various outcome-based classification systems have been employed in the past, the Spitz system is the most widely accepted. The latter is based on birth weight and the presence or absence of major cardiac abnormalities (please refer to table below).

Following surgical repair, the overall incidence of anastomotic leak is 11-21%. Contrast studies will often show a wisp of contrast outside the oesophageal lumen. However, this is not clinically significant, clinically important leak rates are @5%. Approximately 50% will develop an oesophageal stricture requiring dilatation and clearly these patients are more susceptible to respiratory infections. This is due to loss of epithelium and goblet cells around the area of the original fistula as well as ongoing risk of aspiration. Abnormal development of the mesenteric plexus also results in both abnormal peristalsis (dysmotility) and impaired lower oesophageal sphincter tone, predisposing the infant to gastro oesophageal reflux and aspiration.

TOF can recur in up to 9% of patients; anastomotic leaks, oesophageal stenosis and excessive mobilisation during surgery all contribute to risk.

Spitz Classification:

<table>
<thead>
<tr>
<th>GROUP</th>
<th>FEATURE</th>
<th>SURVIVAL (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Birth Weight &gt;1,500g no major cardiac anomalies</td>
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</tr>
<tr>
<td>II</td>
<td>Birth Weight &lt;1,500g or major cardiac anomalies</td>
<td>82</td>
</tr>
<tr>
<td>III</td>
<td>Birth Weight &lt;1,500g and major cardiac anomalies</td>
<td>50</td>
</tr>
</tbody>
</table>

References:
3. Annual Review, Congenital Anomaly Register and Information service 2011
5. Saxena AK; Esophageal Atresia With or Without Tracheoesophageal Fistula, eMedicine, Mar 2010

P Garcia Pulido, J Mistry, S Barr July 2013, to be re-evaluated July 2016