The Second International Oesophageal Atresia Conference
REPORT – A parent support group perspective.

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The Second International OA conference held on 8 & 9 October in Montreal was a very worthwhile experience from a parental support perspective. The main objectives were:

- Gaining an overview of the world's best practice in the nursing, medical and surgical care of neonates, children and adults born with TOF/OA
- To reassure parents of children with TOF/OA that The RCH is well informed in world best practices & care received is of the highest quality.
- Gaining information which will help facilitate the development and provision of a multidisciplinary clinic for patients with TOF/OA here at The RCH.
- Gaining knowledge on the long terms affects of TOF/OA into adolescence & adulthood, to provide a smooth transition from the care provided at The RCH to St Vincent's Hospital, Melbourne.
- Improving parent support at The RCH for families with an infant diagnosed with TOF/OA.

Hearing the different speakers from all over the world, has reinforced the knowledge that the RCH doctors, nurses and their care given, is among the best in the world. As a support group, we can confidently reassure parents of children with TOF/OA that they are receiving the best care available.
RCH is home to the largest OA database in the world, through the help of funds provided by OARA. It is through this much sort after resource, that many research papers have been published. These papers were often referred to throughout the two day conference.

Recent studies in epidemiology, showed that the incidence is still measured at 1 in 3500 TOF OA cases worldwide. However, the hardest part of making credible conclusions is through the lack of major databases around the world. It is for this reason, that it is so important for international collaboration to continue, and taking it one step further to include the sharing of data. In Europe, there is an association (EUROCAT) that was set up in 1979 which holds records for the epidemiologic surveillance of congenital anomalies. It was set up for the reasons of "pooling data, making comparisons, sharing expertise and a joint approach to European public health questions". This register, however, does not include treatments or follow up care. It is with this in mind, that we could possibly extend our Nate Myers database to include some epidemiology data in the future.
In extending the Nate Myers database, it would also be beneficial to include information about the genetics of the patients. In the Netherlands, blood is taken from the patient at the time of clinical procedures and with consent, used for arrays and karyotyping. Blood is also

taken from the parents for similar testing. Their database has >575 OA patients and this includes genetic data.

The discussions about thoracoscopy vs thoracotomy proved to be an extremely popular discussion. With views these days about key-hole surgery as the best way to achieve the same outcome as open surgery with less recovery time. There were many advantages and disadvantages presented for both. The overall conclusions are that for thorascopies, the surgeon and medical team need to be well trained as it is difficult to complete sutures with such a small access point, however it avoids the complications that can be as a result of a thoracotomy such as rib fusion, scoliosis, nerve damage etc. Ultimately it comes down to the individual surgeon and their experience and the experience of their support team.

Long gap OA was very topical with the most common theme of that being an inconsistency of terminology about what is considered a 'long gap'. Some countries measure in terms of the number of vertebrae of the gap or how many centimetres, some simply say, that it is when it is too long for initial repair. Therefore, one of the aims for the next conference is to come up with worldwide terminology. The consensus overall about long gap, was that a true primary repair is always the best option, if possible. So in this case, all possible scenarios should be looked at including growth induction (Foker technique) and intraoperative traction, before looking at replacement options. In contrast to this, there were also discussions of colon and gastric tubes/interposition and from this opinion there is not enough substantial evidence yet to suggest that the child's oesophagus is the best and more research is required. For this to happen, the discussions continued to revert back to the fact that there needs to be worldwide registers that document each of these procedures, their follow up complications and the quality of life of the patients.

Strictures are a common problem with the repair of OA and it was stated in these presentations that there is no evidence to support routine dilatations are beneficial over selective dilatations. They have found that 50% of strictures improve after 6 months, with 30% being persistent. Possible options available to prevent strictures are intralesional steroid injections (ISI) using triacmcinolone acetomide. Also, mitomycin C, which in 1963 was used to prevent scars of surgery, could help prevent the scarring of the oesophagus, thus preventing strictures occurring. Stents were also mentioned as a solution. At this stage there are no world standards in treatment of initial repair, reflux and strictures.

The psychological impact session was extremely valuable from a parent's perspective and shows how important it is for families to seek support and understanding from those who have gone through a similar experience via the support groups. This type of trauma has shown to put parents at risk of depression, and anxiety is very prevalent. There have been studies to show that post traumatic stress disorder can be as a result of having a baby born with major life-threatening conditions such as TOF OA. There were also indications that the

child would develop PTSD and this could come through with troubles with sleeping and new fears & anxiety. This is an area that needs more research to better deal with supporting parents early on, so that PTSD is not as severe months or years later.

The presentation by the family highlighted the importance of doctor referral to support groups as it takes away that 'clutch' of the surgeons and doctors and can help build confidence in parenting these children. Support groups can also help with funding into new research initiatives.

On day two of the conference, the challenges and complications were discussed including those of reflux, dysmotility, feeding difficulties, pulmonary & airway problems and spinal anomalies. This then lead to discussions to the future quality of life and adult follow up.

In presentations by gastroenterologists from Boston and Madrid, the following was reported. Of the patients born with OA, 17-63% of them experience some level of Gastro Oesophageal Reflux Disorder (GORD) and the complications arising from this can lead to failure to thrive, stricture formation, aspiration, dysphagia, chronic lung disease, Barrett's oesophagus and cancer. The obvious treatment of this is through medicines and those that are suggested are acid suppressants, proton pump inhibitors (PPI) and Prokinetic reflux inhibitors, such as Baclofen. Positioning of the body when during sleep can also help and it is said that the prone position is better over the supine position. (Please note: The prone position is not recommended in infants due to the increase risk of SIDS) Also a left lateral position reduces the incidence of reflux, as gastric emptying is found to be faster in this position. However, 27% of OA patients don't respond well to medications and will require surgery, in the form of a fundoplication. It has been concluded that a fundoplication will not necessarily decrease the risk of Barrett's Oesophageal cancer. It is also interesting to note that pH probing doesn't always pick up reflux, as it does not detect non-acid reflux.
Most of the information gathered for these presentations were from research that was a few years old, so it is important to note here that current research needs to be conducted in this area.

Oesophageal dysmotility was discussed and suggested that it is a major component of the OA condition, especially worsened post-operatively and in those with oesophagitis. The best standard for detection, as stated by a paediatrician in Montreal was the oesophageal monometry, which in OA patients, shows a lack in peristaltic wave.
There is currently no evidence to support that the following drugs improve motility; cisapride, domperidone, tegaserod, prucalopride, lincaclotide and metaclopramide. However, the following drugs have had research supporting its use; bethanechol, pyndostigmine and erythromycin. The following herbal drug is also said to be effective, Iberogast. There was also the discussion of a gastric electrical stimulation (GEST) to help with nausea and vomiting.

There needs to be more studies conducted in this area as to overcome this issue.

Feeding difficulties can arise in children born with OA if there have been complications, delayed surgery and therefore delayed introduction of foods, dysmotility, stricture formation and thus repeated dilatation and GORD. Preventative strategies to be used are sham feeding (for delayed surgical OA patients), use of a pacifier to encourage the sensation of sucking, provide pleasurable experiences with food and model feeding behaviours. In conclusion, a multidisciplinary team is required in the following areas, medical, nursing, nutritional and psychological.

Skeletal problems, in particular, thoracogenic scoliosis following surgical correction, can be detected in OA patients and is said to occur in 6-50%. Curves greater than 20 degrees are rare and small curvatures are frequent. The risk factors of thoracogenic scoliosis are frequent surgeries via thoracotomies, and ribs resection. Congenital scoliosis can also be present in OA patients that have the VACTERL association. Common practices to correct scoliosis is through bracing and if necessary, surgery.

A major interest in the medical world of TOF OA is now the follow up of adults and transition into adult care. There is still very little literature on the long-term effects of OA and the research that has been conducted has been with small sample groups. So the areas in need of research are; gastrointestinal morbidity, respiratory morbidity, quality of life and school performance and mental health. Recommendations from this presentation were the need for multi-disciplinary teams for ongoing care, multicentre standardised follow up and international follow up registry.

In conclusion, there is still much research to be conducted in the area of TOF OA and related anomalies. It is the relative rareness of this condition, which means that collaboration around the world is paramount in order to compile enough statistics to make relevant theories and conclusions. It is important that we maintain these international links, so our members benefit from the most up to date research being carried out. The future of these conferences is extremely positive and the 2014 conference is set to take place in Rotterdam, Netherlands. It has been suggested that the 2016 conference be held in Melbourne, Australia which would be extremely beneficial for the RCH and its patients.

It is with this vision in mind that OARA would firstly like to initiate the first National conference for TOF OA, here in Melbourne. Due to the vastness of Australia, it would be impossible to have just one specialist centre for the surgeries of TOF OA, therefore it is so very important that experience and knowledge is shared among the medical profession within Australia, so that the outcome for all TOF OA children Nationally is optimal.