



# **TOWARDS A HOLISTIC MODEL FOR THE TREATMENT OF THOSE BORN WITH OESOPHAGEAL ATRESIA (OA) FROM DIAGNOSIS TO TRANSITION AND ADULT CARE**

## **A TOFS Position Paper September 2021**

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## Foreword

This position paper marks a new direction for TOFS in patient advocacy.

Nearly 80% of our members report raising awareness of OA and OA-related issues amongst medical professionals as our most important service to them, and over 70% also value being able to share experiences with other families affected by OA.

We are extremely grateful to Graham and Julia for preparing this report on our behalf.

Graham is one of the oldest OA survivors, and has been a TOFS Trustee for over 13 years, with a particular interest in developing links with medical professionals and researchers. He was instrumental in setting up the Medical Advisory Group at TOFS; he has presented at the British Association of Paediatric Surgeons (BAPS) conference (in 2015), and in January 2021 Graham was appointed a Patient and Public Voice (PPV) partner in the NHS England Clinical Reference Group (CRG) for Specialised Surgery in Children.

Internationally, Graham was a driving force in the establishment of EAT,<sup>1</sup> where he is now Special Patient Advisor, having been its Chair for the past 10 years. He has also presented at all of the INoEA<sup>2</sup> conferences, and is the lead patient representative in ERNICA.<sup>3</sup>

Through his commitment as a patient advocate, he has established strong working relationships with key medical professionals both in the UK and internationally, and is committed to supporting and encouraging collaboration both with them and, crucially, between them.

Julia is the parent of a child born with OA. In the past four years she has increasingly been involved with TOFS and, in her capacity as a paediatric dietitian, she is a member of our Medical Advisory Group. Julia has written or contributed to several TOFS publications, including our recently launched recipe book, our guide to introducing solids, and – jointly with a speech and language therapist – leading a virtual information session for our members.

Julia presented at the last INoEA conference, and she has helped to develop the ‘CAN EAT’ Care And Nutrition in Esophageal Atresia animation based on the ERNICA OA consensus guidelines. She has also recently become a board member of EAT.

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<sup>1</sup> EAT (Esophageal Atresia Global Support Groups) is the international federation of OA patient support groups: [www.we-are-eat.org](http://www.we-are-eat.org).

<sup>2</sup> INoEA (International Network on Esophageal Atresia) is a multidisciplinary network of healthcare professionals with a particular interest in OA: <http://www.inoea.org>.

<sup>3</sup> ERNICA is the European Reference Network for rare Inherited and Congenital (digestive and gastrointestinal) Anomalies. It is a network of expert multidisciplinary healthcare professionals from specialised healthcare providers across Europe (‘[ERNICA members](#)’). ERNICA has 19 member hospitals across 9 EU member states: <https://ern-ernica.eu/about/ernica/>.

What we hear from parents strongly suggests that there is variation in the paediatric care of those born with OA, and families become acutely aware of this when they share experiences.

We also often hear distressing stories from adults born with OA who simply don't know where to turn, frustrated with the lack of understanding about their condition and associated complications, and who are desperate to know where to find specialist medical support. As it stands, we are aware of some clinicians that have an interest in this area, but this isn't enough to provide adequate care.

We welcome a commitment to lifelong care, and we would support appropriate centralisation of both paediatric and adult services to ensure all those with OA can receive the highest standard of care. We would also welcome collaborative research and the development of a lifetime registry.

As the UK's patient advocacy group for OA, we are keen to be involved in the development of these services.

Duncan Jackson – Chair of the Trustees

Diane Stephens – Chief Executive Officer

## **Endorsement by EAT**

Advocating for the recognition of OA as a lifetime condition has been, and remains, a key objective of EAT. At the same time, EAT is convinced that the quality of surgery, perioperative care and ongoing follow-up are critical to long-term outcomes. However, EAT also contends that this quality of care is best delivered by a centralised healthcare model with a limited number of expert centres that can sustain a suitably experienced team of surgeons, who, along with other clinicians and healthcare professionals, can support multidisciplinary follow-up clinics.

We are delighted to support this initiative of TOFS which – although focused on the UK – contains a commentary and recommendations that we contend are valid internationally.

Anke Widenmann-Grolig, President, EAT      JoAnne Fruithof, Vice-President, EAT

## Acknowledgement

As described below, the position adopted and recommendations made in this paper have been developed over several years, and much of our vision pre-dates the recent NHS England Getting It Right First Time (GIRFT) review into paediatric surgery (which TOFS was invited to comment on).

It seemed timely, therefore, in view of the recent publication of the GIRFT report, to ensure that our observations and concerns as a patient organisation are considered as part of the expected NHS England re-configuration of services. We are extremely grateful to the many clinicians who are generous with their time and expertise in supporting TOFS. We look forward to working together in our joint ambition to support the best outcomes for those born with OA.

## Purpose of this position paper

To ensure that the views of patients with oesophageal atresia (OA)<sup>4</sup> and those of their families are taken into account by NHS England, Health and Social Care Board (Northern Ireland), NHS National Services Scotland and NHS Wales in the development of neonatal services/operational delivery networks and paediatric surgical centres.

To ask the NHS to commit to the development of lifelong services for OA, acknowledging that care during the neonatal and paediatric period is likely to have an effect on outcomes as an adult.

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<sup>4</sup> Throughout this document the term OA is used to describe oesophageal atresia and its associated types including (figures in brackets represent prevalence):

Type A: Oesophageal atresia without tracheoesophageal fistula (8%)

Type B: Oesophageal atresia with a proximal tracheoesophageal fistula (1%)

Type C: Oesophageal atresia with a distal tracheoesophageal fistula (85%)

Type D: Oesophageal atresia with a proximal and a distal tracheoesophageal fistula (1%)

Type E: Tracheoesophageal fistula without atresia (H-fistula) (4%)

## About OA

Oesophageal atresia (OA) is a congenital malformation with approximately 1 in 3,200 births recorded in the UK in 2018.<sup>5,6</sup> There are around 160 newborn cases of OA in England each year.<sup>i</sup>

OA can be part of the presenting features for VACTERL Association. VACTERL is an acronym for a combination of congenital malformations of the **v**ertebrae, **a**nus, **c**ardiac tissue, **t**rachea, **o**esophagus, **r**enal tissue and **l**imbs. This occurred in approximately 1 in 17,000 births in the UK in 2018.<sup>5</sup>

The first successful end-to-end anastomosis repair of OA was reported in 1941; the first successful repair in the UK was in 1947. Mortality has reduced significantly in the intervening years and the focus of care for OA patients has shifted to the treatment of long-term morbidity and quality of life issues.<sup>ii</sup>

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<sup>5</sup> Data from EUROCAT Prevalence Charts and Tables – UK – 2018: [https://eu-rd-platform.jrc.ec.europa.eu/eurocat/eurocat-data/prevalence\\_en](https://eu-rd-platform.jrc.ec.europa.eu/eurocat/eurocat-data/prevalence_en)

<sup>6</sup> TOFS estimates that the UK population of people born with OA is approximately 3,000 children plus in round terms some 6,000 adults. Significant numbers of adults need ongoing care and struggle to find it. The total OA population is similar to the cystic fibrosis population of about 10,700 (UK CF registry 2019).

## About TOFS

TOFS (Tracheo-Oesophageal Fistula Support) is a charitable patient support organisation dedicated to improving the lives of all who were born with OA.

TOFS is the second oldest OA support group internationally, and has been established for nearly 40 years. The majority of parents in the UK who have a child born with OA become members, and total membership including international members is in excess of 2,000.

TOFS provides one-to-one support and a range of information (including a comprehensive book) and other resources to families. TOFS is a recognised source of information for professionals with an interest in OA. Through our Facebook group (which we believe to be the largest such group in the world, currently over 4,000 members) we provide a platform where parents and carers of children born with OA can discuss the problems they face and gain support from each other.

We recognise that some people born with OA continue to need help and support as adults, which we endeavour to provide, and we endorse a second Facebook group, which is specifically for adults and teenagers born with OA.

The TOFS charity also takes an active role in supporting research into the causes and implications of these conditions. We are a founder member of EAT, the international federation of OA support groups, through which we are closely connected with international collaborative work to improve outcomes for those born with OA.

TOFS engages with leading professionals who treat OA in the UK, many of whom are members of our Medical Advisory Group.

As a registered charity, we receive no government funding and rely entirely on members' donations, voluntary donations and other sources of charitable income to fund our activities. Whilst in 2020, the charity received income of some £120,000 (and this has grown greatly over recent years), much of the work of TOFS, especially such engagement (and indeed the preparation of this paper) is undertaken by unpaid volunteers.

## Background to this paper

This position paper is written from a patient perspective, as ‘experts by experience’, and it summarises several years of considering what we, as a patient organisation, feel a more optimal structure of care for those born with OA should look like. We have developed a set of criteria which we would expect centres for paediatric surgery and after-care to comply with, and we have considered how the needs of adult OA patients can be met.

These considerations have been informed by the experiences of our members and their families, both in paediatric and adult services, through informal discussions with healthcare professionals, including members of our Medical Advisory Group, members of professional bodies such as BAPS, through attendance at medical conferences in the UK and internationally, and through presentations and discussions at our own TOFS family conferences.

Our views have also been influenced by our growing engagement with OA-related research studies, including our partnership status with ‘TOAST’,<sup>7</sup> our endorsement of ‘FOOD’<sup>8</sup> and our participation in ‘OCELOT’.<sup>9</sup> In addition, via EAT, we are engaged with international collaborative programmes such as INoEA and ERNICA, and were asked to comment pre-publication on the ESPGHAN-NASPGHAN guidelines.<sup>iii</sup>

It is also important to acknowledge the considerable benefits which TOFS has gained through its membership of EAT. Particular benefits for TOFS (and all EAT’s member associations) have accrued from the ability to exchange experiences, share good practice and patient-oriented information, and develop common approaches to common issues. We have all learnt much about the way OA treatment is organised and delivered in different countries, and the respective strengths and weaknesses.

Without this exchange of ideas and the support from our international colleagues, we might not have had the knowledge or confidence to develop a position paper such as this. Much, if not all, of the vision in this paper emerged from deliberations with our colleagues and is, we believe, shared by all member associations of EAT; many of the recommendations – local/national healthcare models and the constraints of geography notwithstanding – are, we would contend, equally applicable internationally.

Finally, we should note that our growing determination to produce this position paper, and our vision and recommendations that follow, have been specifically shaped by our identification of several themes/issues. Notably, these are: recognition of OA as a lifetime condition; inconsistency in delivery models; absences of an OA Registry and Core Outcome Set; relatively less research and limited data about adults; and opportunities for the use of telemedicine. Further commentary about these themes is outlined in Appendix B.

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<sup>7</sup> TOAST ([Treating Oesophageal Atresia to Prevent Stricture](#)) is an NIHR-funded antacid trial.

<sup>8</sup> FOOD (Fabricating Oesophagus for Digestion) is an NIHR-funded regenerative medicine application for congenital oesophageal malformation (long-gap oesophageal atresia).

<sup>9</sup> OCELOT (**O**esophageal atresia **C**ore **E**outcomes **L**ong**T**erm) is a recently launched Core Outcomes study led by Alder Hey Children’s Hospital.



## Summary of our vision and recommendations

'Patient experience must be valued as much as clinical effectiveness, and patients must be involved in service design and delivery.'<sup>10</sup>

**Our vision for lifetime care and surveillance of OA patients to reduce morbidity can be summarised as follows.**

- As part of the NHS long-term plan, the NHS should become more joined up and coordinated in its care, breaking down traditional barriers between care institutions, teams and funding streams. It recognises that there is currently too much variation in service quality between clinical teams and between different parts of the country.<sup>11</sup> We also feel that this is the case in OA care. The Future Hospital Project showed that developing an ethos of patient participation in co-production/co-design of services was achievable. There needs to be proper investment for patient participation at the start of the design process rather than when projects are already set up.<sup>12</sup> We want to see patient participation in co-design of the redevelopment of paediatric surgical services with NHS England, specifically in OA – not just limited to paediatric surgery but in the coordination of multidisciplinary care throughout the life course of those with OA.
- All children with OA should be treated by one of a set of 'expert centres'<sup>13</sup> adhering to the criteria set out below – with a sufficient caseload to maintain competency, training, and to facilitate research and development, including all aspects of interdisciplinary follow-up care through to 'transition' (no 'discharge').<sup>14</sup> We believe an average of ten OA cases per year for each centre is needed for a sufficient caseload to maintain a sub-specialist rota of surgeons. The 'OA centre' should retain responsibility for the management of the patient, even where local hospitals are used for some aspects of follow-up or emergency care. For lifelong services, we would like to see multidisciplinary 'one stop shop' clinics, and the use of technology to improve communication between specialist services and other care providers.
- Co-development with patients of **lifelong care pathways**, including models of care coordination through NHS operational delivery networks (ODNs), and between ODNs (as centralisation of services means there may not be an OA centre within each ODN). The OA expert centres should maintain and lead the OA care for patients. A 'co-design framework to develop telehealth services'<sup>iv</sup> would enhance this model to support follow-up care, avoid unnecessary travel and enable 'virtual' multidisciplinary clinics (MDCs).

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<sup>10</sup> Future Hospital Commission, Future Hospital: caring for medical patients. London: Royal College of Physicians, 2013.

<sup>11</sup> <https://www.longtermplan.nhs.uk/>.

<sup>12</sup> Frith L, Hepworth L, Lowers V, *et al.* Role of public involvement in the Royal College of Physicians' Future Hospitals healthcare improvement programme: an evaluation. *BMJ Open*. 2019 Sep 12;9(9):e027680.

<sup>13</sup> The GIRFT report recommends an 'indicative' number of ten specialist centres, with three of these being for long gap in NHS England.

<sup>14</sup> We seek to distinguish here between 'discharge' where related to a specific period of hospitalisation, and 'discharge' in the sense of a 'final discharge' from the ongoing care of a specialist centre/specialist clinician. Our contention is that OA patients should never be 'finally discharged' until transition to adult care and/or surveillance.

- Within ten years (preferably five), a long-term Core Outcome Set (COS) and a lifetime Registry<sup>15</sup> for OA will have been implemented. The Registry should gather data on the incidence of Barrett's oesophagus and oesophageal cancer, to help establish risk factors, and refine a surveillance programme accordingly.
- A small number of regional centres should be established for adult care, providing MDCs for follow-up and lifelong surveillance. These would link with paediatric centres (particularly important when looking at long-term outcomes). There is limited research in adults, so we are keen to see adult centres committing to research and linking internationally through organisations such as INoEA. Recognising that there is currently a lack of specialist knowledge of adults born with OA, we would like to work with centres and the NHS in helping to develop adult care and follow-up. We would expect adult care to include the currently accepted practice of regular endoscopies and biopsies for oesophageal surveillance.

**A summary of our specific recommendations includes the following.**

NB: Please see the table in Appendix A for further detail and references.

- Neonatal/surgical/paediatric units should adopt the principles of family-integrated care as set out in the Neonatal Critical Care Review<sup>v</sup>/BLISS<sup>16</sup> Baby Charter.<sup>vi</sup> Parents should be seen as primary carers during all hospital stays and be supported to deliver as much of their baby/child's care as is feasible depending on the baby/child's needs and the family situation. To do this, parents need adequate facilities to be resident. This will benefit the infant's neurodevelopment and lead to greater family cohesion, increased success with breastfeeding and reduced parental stress. Centres should inform families of patient support groups such as TOFS at diagnosis.
- The number of centres performing OA repair and leading follow-up care should be reduced to drive forward quality, research and development, and to support the training of future surgeons and the wider multidisciplinary team (MDT). With the current low volumes per centre/surgeon, it seems as though research has stagnated, as is illustrated by the fact that we are no further forward 18 years after the first described thorascopic repair of OA, as its benefits over open repair remain unproven.
- 'Long-gap' repair should be limited to a small subset of these centres for the same reasons, including developing the expertise of the wider MDT. One example of developing practice to reduce morbidity in 'long-gap' OA would be developing sham feeding with a Replogle tube. This is already practised in several centres outside of the UK and has been shown to reduce the time to full oral feeding post long-gap repair.
- A specialist centre should lead on the development of clear pathways/guidance/support for local centres and primary care. Common areas of concern for our members tend to be

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<sup>15</sup> The following OA registries already exist: French National Esophageal Atresia Register; Turkish Esophageal Atresia; and, at present the only international registry, EUPSA-EAR.

<sup>16</sup> BLISS is the UK charity supporting babies born 'premature or sick'; [www.bliss.org.uk](http://www.bliss.org.uk).

respiratory issues and food bolus obstructions. We feel advice for primary care or local centres on the management of respiratory infections, food bolus obstructions and brief resolved unexplained events (apnoea, colour change, change in muscle tone, choking or gagging) should be developed. Centres should have clear and defined capabilities, and 24/7 availability. We support the GIRFT recommendations with regards to co-location of services, to minimise transfer between centres and interruptions/delays in patient care.

- We recognise variation in care between centres at present. One important area for families is support prior to and after taking their baby home for the first time.
- We would like to see OA-specific training in place for all families, including but not limited to:
  - Basic life support training
  - Choke/food bolus obstruction training
  - Information regarding potential problems and what to do in this situation – stricture, reflux, feeding difficulties, tracheomalacia
  - Single point of contact from specialist centre, eg clinical nurse specialist for consistent communication and access to information
- Research

All UK centres (coordinated by a group such as the BAPS ChUGS<sup>17</sup>) should:

- Engage with TOFS to form a joint UK multicentre research strategy (TOFS will engage with its members to inform the process)
- Engage and collaborate in research, development/outcome measures, and registries at a national level (through ChUGS) and at an international level (through INoEA).<sup>18</sup>
- Consider (and where appropriate) further research into the adoption of advanced surgical techniques. As mentioned in the GIRFT report, the first thoracoscopic repair was performed 18 years ago and we appear no further forward as to whether this is more beneficial for patients.

- Transition and adult care

A comprehensive and coordinated transition programme into adult care should be in place to improve the OA health knowledge and self-management skills of young adults. Adult follow-up is important and a lifelong surveillance programme requires engagement with relevant adult services to establish centres for adult care. At TOFS, we are aware of inadequate/non-integrated care for adult patients with ongoing more severe morbidities and the absence of a surveillance programme for all adults born with OA. Most adults do not have any contact with hospital services beyond childhood, and even when symptomatic one-third of adults do

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<sup>17</sup> BAPS ChUGS – British Association of Paediatric Surgeons – Children’s Upper Gastrointestinal Surgical Network.

<sup>18</sup> Other countries (for instance France) have set up an OA registry and a number of research articles have been generated as a result. UK registries exist for those born with other rare diseases, for example cystic fibrosis, which have helped to improve the standard of care and compare outcomes in relation to treatment regimens.

not seek medical advice. A transition programme needs to inform young adults in a sensitive way of the need for ongoing follow-up, the potential health problems that may occur and the importance of reporting any changes in symptoms.

## Adult OA patients

Whilst we acknowledge that there is a growing recognition of the need for lifetime surveillance and follow-up for those born with OA, much of this paper is focused on paediatric care. We want – quite consciously – to take the opportunity here to emphasise that significant improvements in care are needed for adults born with OA.<sup>19</sup>

At present, there are no specialist clinics for OA patients, though there are some adult clinicians in both the respiratory and gastroenterological disciplines who have developed a reputation for treating adult OA patients with particular morbidities.

Neither a coherent follow-up programme, nor even a surveillance programme for adults born with OA exists in the UK. Whilst we have already remarked that no consistent model of transition is in place at the paediatric surgical centres, the absence of specialist clinics or even of a recognised list of adult clinicians with experience of adults born with OA, renders any transition programme to be of limited effectiveness. In practice, with some exceptions, the best that paediatric consultants can do at transition is to transfer responsibility to the patient's GP.

We want to state quite categorically that we believe specialist MDCs for adults born with OA should be established – preferably as a minimum within NHS England at three locations, covering broadly the North, the Midlands and the South of England. Consideration should also be given by the NHS in the other nations of the UK to establishing one specialist MDC each.

Whilst we recognise that each patient may have specific morbidities, we would recommend that these MDCs cover many of the disciplines associated with paediatric MDCs, including dietetics, gastroenterology, pulmonology, psychology, and speech and language therapy. Furthermore, for young adults at least, we would recommend that the MDCs include on a consultative basis a paediatric surgeon or paediatrician specialising in OA, preferably, where possible, the patient's own consultant.

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<sup>19</sup> We would suggest that plans for the care of adults born with OA should be informed by a review of international best practice. The Netherlands may well be at the forefront, as an audience of OA adults heard at the TOFS 2018 Adult Seminar (Spaander MCW Prevalence of Barrett's Oesophagus and Oesophageal Cancers in oesophageal atresia. A presentation to the TOFS Adult Seminar, June 2018. Video available via the TOFS website). TOFS understands that the Netherlands already has follow-up clinics in place for adult TOF patients, and has offered oesophageal screening to its OA adult population. Some relevant healthcare recommendations have also been published (C.A. ten Kate, A.B. Rietman, L.S. Kamphuis et al., Patient-driven healthcare recommendations for adults with esophageal atresia and their families, *Journal of Pediatric Surgery*, <https://doi.org/10.1016/j.jpedsurg.2020.12.024>).

NHS England has a specification for adult congenital heart disease<sup>vii</sup> and we would welcome something similar for OA, to include that any oesophageal/gastric/bowel surgery required as an adult should be carried out in a specialist tertiary centre. There should be prompt advice sought, and, where appropriate, transfer to a specialist tertiary centre so that adults born with OA can be treated in the most effective and efficient way.

Finally, we want to emphasise that even for adults with no discernible or minimal morbidities, surveillance (eg every five years) would be a highly desirable, preventative practice, where signs of conditions such as Barrett's oesophagus could be identified. Barrett's oesophagus is reported in up to 12.5%<sup>viii</sup> of adults born with OA, whereas it has a prevalence of 1.3–1.6%<sup>ix</sup> in the general population.

We will also continue to advocate for an increased emphasis on research on adults born with OA including 'Quality of Life' studies.

## Final word

As stated in the introductory sections above, this paper is written from the patient perspective; it seeks to address issues about which we have strong opinions as we advocate on behalf of patients and their families for improvements in the structure and scope of the treatments available for those born with OA.

One might imagine that a support group such as TOFS would want to see high-quality expert care for those born with OA at every hospital. TOFS is entirely clear that this cannot be done, simply because of the relatively small numbers of those affected by OA. Once into the post-surgery aftercare phase, most parents of children born with OA quickly learn that it is well worth journeying a long way (more than an hour) to receive care from an expert specialist. As evidenced by the TOFS support groups on Facebook, inadequate generalist healthcare at their local district general hospital or from their GP is a source of frustration amongst parents of children with OA.<sup>20</sup>

Moreover, TOFS is aware of several cases of adults born with OA who have successfully accessed respiratory care with one renowned expert several hundred miles away from their homes.

We believe that our recommendations are practical and achievable and would lead to a more coherent, patient-centric care pathway, one which would not be in conflict with a more sustainable delivery model (a model that we would contend is in many respects applicable to other congenital malformations, such as congenital diaphragmatic hernia and Hirschsprungs disease).

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<sup>20</sup> The desirability of centralising OA repair surgery was eloquently explained by Mr Matthew Jones at the TOFS Conference in 2019. This presentation was well received by the audience, mostly of parents of younger children who were born with OA (video link available on TOFS website).

We also acknowledge the ‘real world’ and recognise that the competition for finance and resources in the NHS often brings constraints on what is possible. Although this is not our focus in this paper, we do believe that our recommendations are compatible with increased cost-effectiveness, whilst providing a framework for improved patient outcomes – the ultimate goal for us all.

We believe that a major opportunity now exists to take a significant step forward in implementing a coherent care pathway for OA patients deployed at a reduced number of ‘expert surgical centres’ – each with a ‘critical mass’ caseload which can deliver sustainability of surgery, leading to improved outcomes by increasingly ‘Getting It Right First Time’. Together with an interdisciplinary approach to follow-up, we also believe that commitments to research and collaboration are vital ‘pieces of the jigsaw’ for centres that can be declared truly expert.

We also see a crucial opportunity to enhance the patient experience through a patient-centric model that exploits new technologies, adopts ‘next practice’ in improved integration of services provided directly by the centres themselves and those provided by local hospitals (including A&E departments). We believe that such a model has the potential to increase both cost-effectiveness and outcomes. Crucially, we are passionate in our advocacy of lifetime surveillance and follow-up, with effective transition to properly established and funded MDCs for adults born with OA.

We suspect that much (though not all) of what we say is actually not that new, and we recognise that our patient perspective, whilst important, is but one which can influence how OA-related healthcare is delivered in the future. We also appreciate that in an organisation as complex as the NHS, there are inevitably some vested interests, which might respond defensively to our observations and recommendations; we also recognise that change is easy to promote and much more difficult to implement. However, all we ask is that our voice is heard, that our opinions are respected, and that – following due consideration – a ‘can do’ mentality prevails. Essentially, we all want the same: the best outcomes for those born with OA.

## Appendix A: Defining an 'expert centre' and coordination of care

It is the contention of TOFS (and EAT), that the implementation of a consistent and more coherent care pathway from diagnosis (whether ante- or post-natal) to transition demands a re-appraisal of how expert centres (ie in the UK, the specialist surgical centres) are defined. There are no apparent (to us at least) criteria on how the existing centres were defined (and there are parallel observations about how centres were established in several other countries).

In the UK, and in particular in England, we note the recommendations from the recently commissioned NHS England GIRFT<sup>i</sup> study into paediatric surgery (in particular those related to the reduction in the number of specialist centres, and the potential role of ODNs). Its commentary about important lessons learnt during the COVID-19 pandemic, together with the patient perspective outlined in this paper, could, and should, act as a catalyst for developing and delivering a patient-centric service that is fit for purpose in the 2020s.

The table that follows pulls together in more detail our collective thoughts, experiences and views, and the evidence we have found regarding the coordination of care throughout the OA patient journey.

Category	Commentary
Antenatal diagnosis of OA	<p>Every effort is made to keep a mother and her baby/babies in the same hospital during their respective admissions.<sup>iv</sup></p> <p>Parents are given the opportunity to accompany their baby and, where this is not possible, alternative transport arrangements are made and parents are offered the opportunity to see their baby prior to transfer.<sup>iv</sup></p>
Parents as primary carers <ul style="list-style-type: none"> <li>- Surgical NICU</li> <li>- All future hospital stays</li> </ul>	<p>Parental inclusion benefits the neurodevelopment of the baby during critical periods of early life and promotes long-term quality of life and family cohesion. Breastfeeding is more successful, bonding is encouraged and parental stress is reduced, all of which have long-term benefits for babies and families.<sup>v</sup></p> <p>Parents should be the primary care provider for their child and thereby facilitated to work with the clinical team to deliver as much cot-side care as is feasible, depending on their baby's need and their family circumstances.<sup>v,vi</sup></p> <p>Parents require support and facilitation by a service that has appropriately focused and trained nursing or AHP staff, working alongside medical and nursing clinical practice staff. Parental support involves education for parents in the specialised needs of their baby and the training of all staff in the provision of developmentally sensitive care from a multidisciplinary team.<sup>vi</sup></p> <p>All of the above follows the principles of family-integrated care, and units signed up to the BLISS Baby Charter have made a commitment to deliver this in their units.</p> <p>Parents need facilities and resources for them to be resident with their baby for some or all of the 24-hour period if their circumstances permit.</p> <p>As per the toolkit for high-quality neonatal services,<sup>iv</sup> parents should have access to accommodation, including resources to prepare and eat meals, and quiet space. Support for travel, car parking etc, is equally important, as without such provision, parents will face further stress to remain with their baby. This is of particular importance for those diagnosed with long-gap OA, as they may remain in a specialist centre for weeks or months.</p> <p>Trusts have policies to provide financial support for families during long-term admission and/or long-distance transfer away from their local unit. Information about this financial support is made available to families.</p> <p>Families are able to easily access psychological and social support and parents are given written information (in appropriate languages and formats) about relevant services covering at least:<sup>v</sup></p> <ul style="list-style-type: none"> <li>• Local and national support groups (TOFS/EAT)</li> <li>• Social services</li> <li>• Spiritual support</li> <li>• Counselling</li> <li>• Benefits advice</li> </ul>



	<ul style="list-style-type: none"> <li>• Availability of other support services (for example, help with care of other children).</li> </ul> <p>Parents/carers of children with OA have been shown to have higher levels of anxiety and depression compared to parents/carers of children with chronic health conditions. Parents/carers more likely to be at risk of poorer mental health outcomes were younger, may have financial concerns and reported OA-related feeding problems.<sup>x</sup></p>
<p>Reduction in number of OA surgical centres</p>	<p>As per the GIRFT report<sup>i</sup> into paediatric surgery, the current number of centres performing OA repair should be reduced to support sufficient repairs per centre and surgeon to maintain competency.</p> <p>There is a consensus recommendation that OA centres should have a minimum average of five new OA cases per year. OA patients should be operated on and treated in specialist OA centres, with an MDT follow-up and transition.<sup>xi</sup> At the time of this consensus statement, patient organisations were asking for a minimum of ten average cases per year and we believe that this is needed to maintain a sub-specialist surgical rota.</p> <p>Long-gap OA should be managed in centres with expertise in oesophageal reconstructive surgery. These centres should have more than two cases per year.<sup>xii</sup> At the time of consensus, patient organisations were asking for a minimum of five cases of long-gap OA per year.</p> <p>Within adult care, the Association of Upper Gastrointestinal Surgeons (AUGIS) have recommended that individual specialist surgeons undertaking oesophageal and gastric cancer resections should carry out a minimum of 15 to 20 resections per year, working within a trust comprising six to eight surgeons. This led to a centralisation of surgery, resulting in improved outcomes for these conditions and emergency upper gastrointestinal surgery.<sup>xiii</sup></p> <p>The GIRFT report<sup>i</sup> indicated large variance in the number of OA cases treated per specialist trust and surgeon.</p> <ul style="list-style-type: none"> <li>• Trusts are treating between 1.4 and 18.5 cases per year.</li> <li>• The national average for cases per surgeon is just 1.4, whilst the busiest surgeons perform an average of 3 cases per year.</li> <li>• It is concerning that 40 surgeons claimed to have competency in oesophageal replacement surgery, which at 16 cases per year works out as an average of 0.4 cases per surgeon per year.</li> </ul> <p>As per the GIRFT report,<sup>i</sup> these low volumes make training, research and innovation extremely difficult.</p> <p>As patient representatives, this is frustrating, as we are aware of how important it is to have research/developments focused to reduce the long-term morbidity of this condition. With such low volumes per centre/surgeon, it feels like research</p>

has stagnated, as is illustrated by the fact that we are no further forward 18 years from the first described thoracoscopic repair of OA, as its benefits over open repair remain unproven.

Variability between and within OA centres in the diagnostic work up of neonates with OA has recently been reported from the International EUPSA-Esophageal Atresia Registry.<sup>xiv</sup> Interestingly, this involved 23 centres with numbers of patients per centre ranging from 1 to 49 over a three-year period. We would hope that a reduction in the number of centres would lead to standardisation of care to what is considered best practice. This also highlights the importance of communication and collaboration within centres.

We also welcome the statement in the GIRFT report that ‘successful outcomes and the ability of children born with major congenital malformation to achieve their lifetime potential is also dependent on the experience and availability of all relevant clinicians and allied health professionals (AHPs),<sup>i</sup> in particular ‘children born with VACTERL association, who need input from many different clinicians and AHPs’.<sup>i</sup>

To drive forward quality, research and development, and to support the training of future surgeons and the wider MDT, we agree that there should be a reduction in the number of centres carrying out OA repair and leading follow-up care. Within each centre, we would also like to see the sub-specialisation of surgeons to increase the caseload per surgeon to also benefit research, development and training.

Within the reduced number of centres treating OA, we would like to see the repair of ‘long-gap’ OA limited to a small subset of ‘centres of excellence’. This is because around 10% of OA cases are long gap, so it makes sense to concentrate these cases in fewer centres that can be more proficient and develop expertise/research in oesophageal replacement and the care of these babies. This will involve developing the expertise/skills of the wider MDT. One example would be developing sham feeding using a Replogle tube, which in several centres outside of the UK has been shown to reduce the time to full oral feeding post long-gap repair.<sup>xv,xvi,xvii</sup>

Without dedicated ‘centres of excellence’ for the treatment of long-gap OA, protocols and expertise as above are unlikely to be developed. As patient representatives, this is frustrating when we can see developments happening outside of the UK which seem to reduce morbidity. For the wider NHS, reducing morbidity has the potential to reduce the costs of ongoing care for these patients.

The ERNICA consensus statements provide some guidance on the number of cases per centre; however, even these appear to be low numbers.

<p>What does an OA centre look like?</p>	<p>An OA centre will co-produce services and pathways with patients, patient representatives such as TOFS, and carers, to better understand the challenges and opportunities they face and to create more effective health services.<sup>xviii</sup></p> <p>It is likely that ‘no one size fits all’ for centres, but commitment is required from centres to look at the best fit for their particular centre, their geographical location and service users. Models of care from other specialities, such as cardiology and cystic fibrosis, should be considered. Consideration should be given as to how this may work for OA, such as hub-and-spoke models with local services, which have clearly defined capabilities.</p>
<p>What does a complex care/long-gap OA centre look like?</p>	<p>Centres should ‘use a co-design framework to develop telehealth services to support individualisation of care and meet patient/parent needs’.<sup>iv</sup> This could enable delivering specialist centre clinics to work jointly with local specialists to maintain/develop local team knowledge.</p>
<p>Care coordination</p>	<p>As the NHS long-term plan states, ‘the NHS will increasingly be more joined-up and coordinated in its care’.<sup>xix</sup> In rare conditions, it has been recognised that ‘coordination of care should be family-centred, holistic (including a patient’s medical, psychosocial, educational, and vocational needs), evidence based, with equal access to coordinated care irrespective of diagnosis, patient circumstances, and geographical location’.<sup>xx</sup></p> <p>Patients with rare conditions have reported a lack of support and information provision for rare diseases. Equally, healthcare professionals working with those with rare conditions reported needing support in accessing specialist knowledge. This highlights the importance of training and education for care providers.</p> <p>Patients have reported specialist/condition-specific clinics as a useful method of coordination. Barriers to specialist clinics include funding and patients needing a way to assess whether a centre is in fact a centre of excellence.<sup>xxi</sup></p> <p>Patients report a lack of communication between care providers and that they end up acting as a medium for sharing documents between providers. They report message fatigue, repeating the same information multiple times, as healthcare professionals don’t seem to talk to one another.<sup>xxi</sup></p> <p>Technology may be a useful way to improve communication between healthcare providers.</p> <p>Interestingly, rare disease patients have identified that to promote quality care and coordination, centralised and shared care pathways are needed. Patients reported being happy to travel to access care, if it meant they received expert care.</p> <p>Patients with rare diseases report having to attend multiple appointments, on different days, gaps and delays in sharing of documents, and disagreements</p>

between healthcare professionals. They also expressed concerns regarding coordination of care in emergency situations and the importance of patients taking control themselves in order to mitigate worries.<sup>xxi</sup>

This is important in OA for emergency presentation with food bolus obstruction. This is also recognised as an issue for eosinophilic oesophagitis, and the patient support group for the EOS Network has produced a patient-held information sheet.<sup>xxi</sup> This has a care plan for an emergency admission.

Adult guidelines recommend rapid endoscopy for the removal of oesophageal obstruction within two, or at the latest six, hours when there is complete obstruction (unable to swallow saliva), and urgent within 24 hours when there is partial oesophageal obstruction.<sup>xxii,xxiii</sup>

We recommend something similar should be considered for OA patients with an agreed protocol nationally between centres.

Another aspect particularly related to OA patients is radiation exposure; frequent radiological examinations result in an increased malignancy risk. Care coordination and standard care protocols should be developed to minimise radiation exposure, ensuring all care settings are aware of this.<sup>xxiv</sup>

As discussed above, a reduction in the number of OA centres, to enable clear and defined capabilities and competence, including a caseload that can guarantee experienced clinicians, training and mentoring of more junior clinicians, plus 24 x 365 availability, are all imperative.

All patients born with OA should be treated at a specialist centre with an MDT of different clinical professionals with OA expertise.<sup>xii</sup> Follow-up support and/or surveillance should be both lifelong and structured and led by a centre of excellence. This is necessary to monitor and treat any difficulties that may arise during the life course, such as breathing, nutritional or digestive issues.

Centres should:

- Ensure 'no discharge', and ongoing surveillance of all patients including those with no discernible ongoing morbidities
- Inform parents of children born with OA about parent and patient support groups (TOFS) as early as possible. Centres should recognise patient support group engagement as a beneficial and normal aspect of all healthcare initiatives (co-production of research, patient information/resources, improvement projects etc). Patient organisations and rare disease charities are perceived to be key in care coordination, as they support patients and carers to develop expertise and take control of their condition

	<ul style="list-style-type: none"> <li>- Provide a single point of contact within specialist multidisciplinary specialist services, for example, clinical nurse specialist for patients and local centres to access advice/support</li> <li>- Engage with, and signpost to, third sector organisations (TOFS) to deliver disease-specific information</li> <li>- Invest in technology to support assessment at home</li> <li>- Work to maintain community healthcare services even during periods of high resource need, wherever possible</li> <li>- Acknowledge the burden of parental decision-making during routine follow-up appointments</li> <li>- Identify parental anxiety/mental health concerns related to child’s health/development, and signpost for appropriate support.</li> </ul> <p>All UK centres in conjunction with the BAPS ChUGS should:</p> <ul style="list-style-type: none"> <li>- Engage with TOFS to form a joint UK OA multicentre research strategy. TOFS will engage with its members to inform the process</li> <li>- Be involved in research &amp; development/core outcome measures,<sup>xxv</sup> and registries at a national level (with ChUGS) and at an international level (through INoEA).</li> </ul> <p>We would specifically recommend research into the adoption of advanced surgical techniques (eg endoscopic surgery/minimally invasive surgery (MIS)). As mentioned in the GIRFT report, the first thoracoscopic repair was performed 18 years ago and we are no further forward regarding whether it is more beneficial for patients. With centralisation of complex/long-gap OA centres, the larger caseload through these centres is likely to support research into this.</p>
Research	<p>All centres should:</p> <ul style="list-style-type: none"> <li>- Engage with relevant initiatives of professional bodies such as BAPS and ChUGS, and develop linked groups within other professions, nationally and internationally, eg AHPs, specialist nurses to support professional development and practice sharing</li> <li>- Develop outcome monitoring/measurement to include Patient Reported Outcome Measures (PROMs) and Patient Reported Experience Measures (PREMs) with TOFS/EAT, including these measurements in adults and continuity in Quality of Life (QoL) measures from childhood through to adulthood.</li> </ul> <p>We would like to see research into the incidence of Barrett’s oesophagus and oesophageal cancer in adult OA survivors, along with the identification of risk factors to enhance/inform surveillance programmes.</p>
Home – from specialist centre/to local centre	<p>Following the principles in the toolkit for high-quality neonatal services<sup>x</sup> regarding discharge home:</p>

Parents should be involved in multidisciplinary discharge planning from the point of admission and plans should be continually reviewed.

Families should have appropriate information and training before being discharged home. **OA-specific training at discharge should include:**

- Resus training
- Choke/food bolus obstruction training
- Information and discussion re potential problems: – stricture, reflux, feeding difficulties, tracheomalacia
- Information on who/how to contact in case of suspected OA problems
- Being provided with a single point of contact from specialist centre MDT, eg clinical nurse specialist for consistent communication, access to information.

**Note:** Severe tracheomalacia is typically clinically evident from birth, but children may not show symptoms before 2-3 months of age.<sup>xxvi</sup>

Intermittent compression of a malacic trachea during bolus progression in the oesophagus can cause desaturation, leading to poor feeding and consequently poor weight gain.

Tracheal obstruction can cause ‘apnoeic spells’ or ‘blue spells’. Potentially, this can be due to a bolus of food in the oesophagus compressing the trachea.<sup>xxi</sup>

The baby and family should have their ongoing needs at home coordinated and met by health professionals appropriately skilled in delivering OA care and support in the community, eg members of the centre’s MDT/Clinical Nurse Specialist (CNS).

Centres should identify parental anxiety/mental health concerns related to child’s health/development, and signpost for appropriate support.<sup>xxvii</sup>

Developing transition and adult care

A comprehensive and coordinated transition programme into adult follow-up and/or lifelong surveillance<sup>xxviii</sup> requires engagement with a relevant adult service to establish centres for adult care. At TOFS we are aware of inadequate/non-integrated care for adult patients with ongoing more severe morbidities and the absence of a surveillance programme for all adult OA patients.

Most adults do not have any follow-up, nor do they have any contact with hospital services beyond childhood, and it is reported that even when symptomatic, one-third of adults don't seek medical advice.<sup>xxix</sup>

This, coupled with the increased incidence of Barrett's oesophagus and oesophageal cancer, highlights why lifelong care and surveillance endoscopies every five-to-ten years to detect oesophageal changes are important.<sup>iii</sup> This would enable treatment to start early as indicated, and prevent the development of oesophageal malignancy.

Barrett's oesophagus is considered a premalignant lesion in the general population which predisposes to oesophageal cancer.

Barrett's oesophagus is reported in up to 12.5%<sup>viii</sup> of adult OA patients, whereas in the general population it has a prevalence of 1.3–1.6%.<sup>ix</sup>

Barrett's also occurs at a much younger age in OA survivors, with a median age at diagnosis of 37 years as opposed to that of the general population of 57 years.

To date, there have been 11 cases reported of oesophageal cancer in adult OA survivors, three cases of oesophageal adenocarcinoma, and eight cases of oesophageal squamous cell carcinoma (most found at or near the site of anastomosis).

The pathogenesis of oesophageal squamous cell carcinoma (SCC) may be similar to achalasia, where it is thought to result from stasis, resulting in bacterial overgrowth with nitrosamine production, subsequent oesophageal inflammation and eventually cancer.<sup>viii</sup>

These have all occurred at a relatively young age, with the youngest at 20 years of age.

This highlights the importance of surveillance programmes and gathering data on incidence.

Large prospective follow-up cohort studies are needed, to define the risk and incidence of Barrett's oesophagus and cancer in adult OA survivors. This should lead to the identification of risk factors to refine surveillance programmes, making them more beneficial to patients and cost-effective in the longer term.

A recent systematic review into Barrett's oesophagus and oesophageal cancer following OA repair<sup>xxx</sup> summarised that the lack of documented progression of Barrett's to oesophageal cancer in patients born with OA currently means that the importance of gastric or intestinal epithelial metaplasia in this population cannot be evaluated.

The higher incidence of oesophageal SCC in patients born with OA also warrants further research. At present there are no recognisable precursor cells for SCC, so at present endoscopic surveillance based on Barrett's would not be effective. The review concludes that 'international collaborative studies should be undertaken to identify the optimal screening and surveillance programmes in this population and assess their clinical benefit and cost-effectiveness'.<sup>xxxii</sup>

We are also aware of the development of gastric-neuroendocrine tumours (g-NETs) as a complication of proton pump inhibitor (PPI) therapy. Although NETS are rare in occurrence and slow to develop, their incidence is likely underestimated at present.<sup>xxxii</sup> In OA patients who may have been on PPIs from an early age for prolonged periods, and may have periods throughout their life course on PPIs, we worry that they may be at increased risk of developing NETS. This again emphasises the importance of an international registry for measuring long-term outcomes to evaluate these potential late effects of treatments. This will help patients, families and clinicians together to evaluate the potential risks/benefits of treatments and make informed, shared decisions regarding treatment options.



## **Appendix B: Some key themes/issues noted during the development of this paper**

### **OA is a lifelong condition**

We note that care pathways have been transformed over time from an (almost sole) focus on surgical repair (with parents advised that ‘the problem is fixed’) to a regime that recognises and implements the need for ongoing multidisciplinary follow-up. We do, of course, recognise that a high-quality outcome is highly unlikely without high-quality surgery and perioperative care, but we welcome the implementation of follow-up care pathways involving a range of non-surgical disciplines. We also welcome the declarations of support for ongoing after-care, surveillance and follow-up into adulthood.

### **Inconsistency in delivery models**

The above notwithstanding, our experience as patients indicates that there is no consistent care pathway across the existing surgical centres in the UK, and there is considerable variability in, for example, the existence of MDCs, discharge protocols, coordination with local (non-specialist) hospitals, and no procedures for transition from paediatric to adult care. Transition ‘is not a given’, and these preceding statements are all exacerbated by limited awareness of OA by GPs and in A&E departments.

### **Absences of an OA Registry and Core Outcome Set (COS)**

Despite the significant amount of research carried out into OA over the past years, we note the absence of an agreed and implemented COS. We believe that such a COS should be developed with patients to enhance our collective understanding of the outcomes affecting OA patients, and hope that progress might be made via the recently launched study in the UK (OCELOT), and a complementary registry initiative being undertaken by ERNICA.

### **Relatively less research and limited data about adults**

We note a relatively lower amount of research into adult OA patients. There have been studies which have indicated the enhanced risk of conditions such as Barrett’s oesophagus, which have provided evidence to support the concept of lifelong surveillance, and other studies which have indicated the relatively high prevalence of ongoing morbidities, such as GI reflux and respiratory infections. However, there have been few studies to review a significant cohort to determine the range of outcomes experienced by adult OA patients.

### **Opportunities for the use of telemedicine**

Increased use of information technologies is a growing feature of modern healthcare. By way of example, we note the CPMS (clinical patient management system) developed by the EU for use by the European Reference Networks (ERNs) for facilitating expert panels on complex cases, and regret that, following ‘Brexit’, this facility is not available for UK-based clinicians and hence UK-based patients. We also note the increased use of remote consultations – driven in the past 18 months in response to COVID-19.

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