

E02/S(HSS)c

**2013/14 NHS STANDARD CONTRACT
COMPLEX TRACHEAL DISEASE SERVICE (CHILDREN)**

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – Service Specification

Service Specification No.	E02/S(HSS)c
Service	Complex tracheal disease service (Children)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

The service deals with the management of children with serious tracheal disease in childhood. It is primarily concerned with the treatment of long segment congenital tracheal stenosis (and its associated [60%] lesions), severe tracheo-bronchomalacia and a variety of other, rarer pathologies. Management involves assessment of airway disease by bronchoscopy, bronchography, optical coherence tomography, echocardiography, 3-D imaging by **computed tomography (CT) and** magnetic resonance imaging (MRI). Treatment may involve surgery, for example slide tracheoplasty for tracheal stenosis, or stenting for malacia. Follow up is both by shared care with referring institutions but by annual review at Great Ormond Street Hospital for Children NHS Foundation Trust (GOSH) with anatomic, physiologic and quality of life assessment.

Evidence base

Long segment congenital tracheal stenosis.

The management of this condition has evolved considerably during the last 20 years, before which mortality and morbidity were unacceptably high. During the 1990's most surgery involved the use of patches of various kinds. The long-term results of this approach were, in most hands, disappointing and numbers reported small. Slide tracheoplasty has become the treatment of choice in the 2000's. During the last decade, continued improvements in the patient pathway, and lengths of stay,

ventilation and intensive care unit (ICU) stay have all been reduced. Late mortality has been very low and late morbidity minimal other than regular review. The ages of the children have limited our ability to undertake detailed physiologic studies, but they are now approaching ages when this can also be part of their follow up. Currently, there are no long term quality of life measures in the literature, but the provider is developing an adapted tool from the palliative care quality of life instrument (pcqli) inventory scoring system to describe quality of life with accuracy.

The provider has recently introduced tracheal stem-cell transplantation (British Medical Journal 2010;340:c1633), and believe that this advance will permit correction of severe airway disease currently unamenable to surgery, it provides excellent fall-back options for children with recurrent stenosis or stent-related complications.

Stenting

Airway stenting has been controversial for many years. There has been no ideal stent, and to date each has had important and stent-specific complications, including erosion into nearby vessels, granulation tissue formation, obstruction to secretion clearance and constrained growth. The provider has developed considerable expertise in stenting in children and infants. A recent study has been started for absorbable stents that offer considerable potential advantages in children in that they could deal with temporary malacia or stenosis without inhibiting growth or leaving behind foreign tissue. Initial trials are encouraging*.

Management of tracheomalacia

Tracheobronchomalacia represents a significant burden to the health service and to affected children and their families. It has traditionally required long-term ventilation, either at hospital or at home, and often tracheostomy. With a combination of surgery by aortopexy, stenting as required and rare use of tracheostomy the provider has greatly improved the options for these children, and suggests that earlier referral and treatment might reduce costs to the NHS. Absorbable stents will clearly have a significant role (see above). The advances in stem-cell therapy it is hoped to add new therapeutic options to this difficult group.

2. Scope

2.1 Aims and objectives of service

The service promotes appropriate referrals from across England.

This service is devoted to the management of children with serious tracheal disease in childhood. It is primarily concerned with the treatment of long segment congenital tracheal stenosis (and its associated [60%] lesions), severe tracheo-bronchomalacia and a variety of other, rarer pathologies. The service aims to be the world's leading

centre for such treatment, both in terms of volume of practice and quality of service and outcomes. It aims to be innovative, evidence-based and open in its reporting. The true incidence of these lesions remains unclear, but they are all rare. It has been demonstrated over the last four years that the service can deal with the entire UK population of patients with these conditions, although milder forms of malacia are best treated at host institutions.

The care of these patients requires intense teamwork and considerable experience. It is the view of the service that there is no place for occasional practice, and the service wishes to spread this message widely in the paediatric community.

Objectives

Strategic objectives

- to provide an exemplary and comprehensive service for all eligible referred complex tracheal patients
- expert diagnosis utilising the most up-to-date validated diagnostic tools and knowledge
- expert management of patients through the use of the most up-to-date clinical protocols to achieve the lowest international mortality and morbidity in this field
- clinically appropriate consideration and provision of surgery within the complex tracheal patient pathway
- effective monitoring of patients
- to provide care with a patient and family centred focus
- to be seen as the leading clinical service and source of expert advice for the diagnosis and management of complex tracheal within the NHS. To do this at the lowest cost and with the maximum patient and referrer satisfaction
- to support local healthcare providers to manage patients with a complex tracheal diagnosis whenever it is clinically appropriate and safe to do so
- provide high quality information for patients, families and carers in appropriate and accessible formats and mediums
- to develop the experience, knowledge and skills of the multidisciplinary team (MDT) to ensure high quality sustainable provision.

Outcomes

- To optimise patient outcomes through careful clinical management and support.

2.2 Service description/care pathway

Referral

Patients will be referred to the service initially by telephone or email from an intensive care unit (ITU) and respiratory consultants nationally and internationally. This will be followed by a referral letter and images, including a bronchoscopy, bronchogram and computerised tomography (CT) scan. All complex tracheal referrals will be scrutinised to ensure that they are compatible with the specified service before a referral is accepted. If a referral appears to be outside the scope of the national service as specified herein, the service should seek clarification from the referrer.

Initial assessment

In most cases it is necessary to repeat the diagnostic investigations, particularly CT, bronchoscopy, bronchography & optimal coherence tomography (OCT), unless they have already been assessed by another known referrer, because they are only performed at the provider or require specific expertise. It is anticipated to do this for all potential tracheal reconstructions (some patients do not require surgery following assessment).

Admission

Where the patient requires treatment, they are classified as either urgent or non-urgent. Treatment will either be surgery or stenting (by interventional radiology). If a patient does not require treatment at this stage they are referred back to the referrer, to manage the condition and re-refer the patient should the condition of the patient worsen. The associated costs of the patient outside of the provider, whilst under the care of the referring hospital are outside the scope of this specification.

Condition classification

The provider treats Tracheal patients whose condition falls into one of three categories with the stated probably treatment strategy of:

- **Very complex**
Clinically this will be a patient episode involving a tracheal reconstruction using slide tracheoplasty or patch +/- additional cardiovascular surgery using cardiopulmonary bypass. The majority of such episodes involved initial surgery with an ICU length of stay of 10 days or more; average 21 days and 11 days on other wards. These patients have required higher intensity in terms of number and duration of stays in follow up episodes to date. The average number of follow ups for very complex patients is approximately seven with an average length of stay in ICU of two days and four days in other wards. As this service is not mature it is possible that the average follow ups per patient may increase.
- **Complex**
Clinically this will be an episode involving surgery that requires an anterior aortopexy +/- other procedures (e.g. nuss bar insertion, sternal enlargement, or other cardiac surgery). Resource utilisation of ICU bed days for complex is two days on average and 3.35 days for the duration of the episode outside of ICU.
- **Simple**
Any other tracheal surgery for children satisfying the service criteria based on their condition. This is more costly than "complex" due to the use of stents. The procedure itself is more straightforward for the patient but the direct non-pay costs are higher. Average stays in ICU and other wards are 1.5 and four days respectively.

These categories should be reviewed and revised as new evidence emerges.

Discharge

Patients are normally discharged into the care of the referring hospital, or for simple stent patients, they may be discharged to home.

Criteria for discharge from inpatient care:

- satisfactory stability achieved with or without medication
- no adverse outcomes anticipated
- no further investigation required
- clinically appropriate arrangements for local care and follow-up have been discussed and agreed by all relevant parties
- parents/carers have demonstrated competence in any care they will be required to provide
- parents/carers understand and have the necessary information to contact the complex tracheal disease service.

All discharge planning will be managed through the MDT with local health and social care providers being fully informed of the patient's condition and any responsibilities they will have to assume. The general principle applied by the service is that patients are referred back to their host institution for planned, shared care as soon as is practicable. This varies from referrer to referrer and is often constrained by ICU, high dependence unit (HDU) or transport availability. Each patient is managed against their own care pathway and communication of this rests with the lead nurse for the service or their deputy. The ability to deliver shared care depends upon the skill and facilities of local services and is thus highly variable.

The service is entirely delivered by Great Ormond Street Hospital for Children NHS Foundation Trust (GOSH). None of the patients have yet reached adulthood; however a joint paediatric-adult airway service across the University College London partner's site has been established.

Follow up

Follow-up work is similar for simple and complex cases. The very complex cases require more frequent follow-up than the complex cases especially in the first two years post-operatively. Complex and very complex cases may be followed up at two and four weeks. All patients will be followed up at three and six months and the stent patients will be followed up at six month intervals thereafter with the other patients at 12 month intervals until they reach transition. The current service model provides follow up services on a day case basis. Patients who live in close proximity to the provider can be seen on an outpatient basis

Some patients are seen as outpatients in the ear, nose and throat and respiratory clinics but this activity is not undertaken as part of the tracheal referral. Annual lung function tests are also done but again, separately from the tracheal referral. The majority of patients will require at least one day case admission within an 18 month period.

Currently shared care arrangements for follow up activity are in place with Birmingham Children's Hospital NHS Foundation Trust, Central Manchester University Hospitals NHS Foundation Trust and Alder Hey Children's NHS Foundation Trust.

The provider is looking to develop this model with other centres wherever possible, as it is convenient for patients. However this arrangement has only occurred when the local hospital is willing and able to undertake shared care arrangements and even then it is only partial. The provider still continues to review the child on an annual and hoc basis, in addition to reviewing all images from the local centres at our weekly meetings.

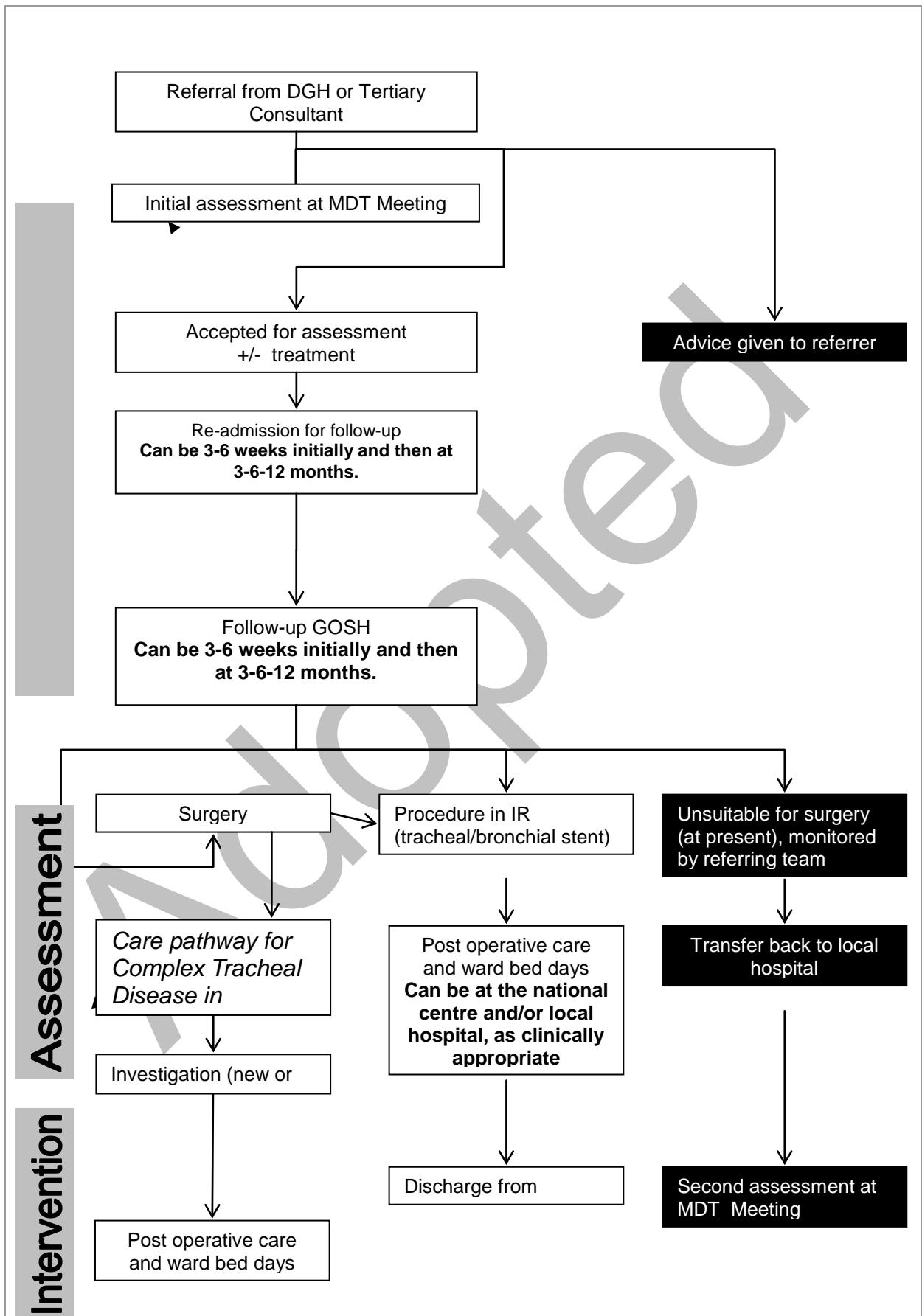
The provider is dependent on these careful long term arrangements fully to inform new patients and their families of outcomes. The emerging evidence base and research demands an ability to study these patients as they grow, at defined intervals.

Each family receives a detailed individualised care plan, and much effort is put into communication of this plan, which is shared regularly via mail. Regular review of progress is carried out at a weekly multi-disciplinary meeting, following which updates to plan are widely distributed.

The provider is developing a patient held record within the cardio-respiratory service, and the tracheal service will be part of this process.

Parents of patients, especially those who are not treated, are encouraged to report any signs of respiratory distress in their child.

Adopted



2.3 Population covered

NHS England commissions the service for the population of England. Commissioning on behalf of other devolved administrations is reviewed annually, and a current list is available from NHSE commissioners.

At the moment the NHS England contract includes provision for the service to treat eligible overseas patients under S2 [Under EU regulations, patients can be referred for state funded treatment to another European Economic Area (EEA) member state or Switzerland, under the form S2 (for EU member states) or the form E112 (for Iceland, Norway, Liechtenstein and Switzerland)] referral arrangements. Providers are reimbursed for appropriately referred and recorded activity as part of NHSE contract.

Trusts performing procedures on EU-based patients outside of S2 arrangements will need to continue to make the financial arrangements directly with the governments involved, separately from their contract with NHS England.

With regard to S2, the mechanism for recovery of costs has been via the Department for Work and Pensions overseas healthcare team. They are responsible for agreeing reconciliation and recovery of costs with European administrations. These arrangements were implemented in October 2009, though a similar process existed previously. The financial flows are therefore back into the Treasury rather than back to trusts.

2.4 Any acceptance and exclusion criteria

Acceptance criteria

Referrals are made either directly to the tracheal service administrator or to members of the multi-disciplinary team. Referrals usually come from tertiary cardiothoracic, paediatric ear, nose and throat or general surgical centres. Suitability is judged by the receiving physician, and must fall into one of the diagnostic groups indicated in this specification. The provider does not receive referrals from primary care.

Many patients are too sick to travel without the use of specialist retrieval services to collect the patients as soon as an Intensive Care bed is available.

The provider has a duty to co-operate with the commissioner in undertaking equality impact assessments as a requirement of race, gender, sexual orientation, religion and disability equality legislation.

Although there is an upper age limit of 16 for this service, most patients are referred within their first year of life because of the nature of the disease. More than 10% of patients are on extracorporeal membrane oxygenation (ECMO) when referred and remain so until surgery.

Exclusion criteria

The only exclusion to this service are patients aged 16 or over (unless with relevant other developmental factors allowing consideration of an older age), and those with such severe co-morbid conditions that treatment is not considered viable.

The team has to restrict access solely because of availability of ICU beds, and will expect referring units to 'hold' patients until a bed is available, unless the patient is judged to be an emergency. Priority will be given to those with life-threatening problems. Such patients form a significant minority of patients, and approximately 10% of those with stenosis need cardio-respiratory support with extracorporeal membrane oxygenation (ECMO).

2.5 Interdependencies with other services

Patients are prioritised for admission by the tracheal team at its weekly meeting, after daily review with the clinical lead and at the daily bed planning meeting with the ICU team. Prioritisation has to take place against the needs of the other NHS England services and in the wider context of acute cardiac care. Careful communication with referrers and families is maintained via medical, administrative and nursing staff.

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

This service is fully integrated into the trust's corporate and clinical governance arrangements.

Within the team, all unusual events are reviewed at a weekly meeting, and relevant data is presented regularly at international meetings for benchmarking. A clinical database records complications and the intention is to continue to reduce their incidence. All major events are subject to wider departmental scrutiny and presented a separate, weekly, clinical governance meeting.

See also NHS England Service Standards for the complex tracheal disease in children services.

4. Key Service Outcomes

The outcomes for this service have few international comparators. The centre produces a comprehensive outcome report, of which the measures below are a part.

<i>Quality Performance Indicator examples</i>	<i>Threshold</i>	<i>Method of measurement</i>	<i>Consequence of breach</i>	<i>Report Due</i>
Survival Rates	Historic average	Service review report	Review and, if appropriate, action plan	Annual
No. of stents per Slide Tracheoplasty	Historic average	Service review report	Review and, if appropriate, action plan	Annual
Post-operative Complications (incl. infections)	Historic average	Service review report	Review and, if appropriate, action plan	Annual
Length of stay	Historic average	Service review report	Review and, if appropriate, action plan	Annual
Re-admissions	Historic average	Service review report	Review and, if appropriate, action plan	Annual
Re-referrals by referring hospitals	Historic average	Service review report	Review and, if appropriate, action plan	Annual

5. Location of Provider Premises

There are no formal sub-contractors within this service specification. Informal follow-up links do exist with Manchester, Birmingham and Liverpool children's hospitals. However, these are not formally funded or included within this specification.

Provider
Great Ormond Street Hospital for Children NHS Foundation Trust