

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No:	170051S
Service	Auditory brainstem implant (ABI) for children with congenital abnormalities of the auditory nerves or cochleae
Commissioner Lead	Nicola Symes, Commissioning Manager, Highly Specialised Services & Lead Commissioner, Specialised Ear and Ophthalmology CRG
Provider Lead	<i>For local completion</i>

1. Scope
<p>1.1 Prescribed specialised service This service specification covers the provision of auditory brainstem implants (ABIs) for children as identified in Section 108: Specialist ear, nose and throat services for children in the Prescribed Specialised Services Manual (the Manual).</p> <p>1.2 Description The auditory brainstem implantation (ABI) service is for children with profound deafness who have no functional hearing as a result of congenital abnormalities affecting the auditory nerve or the cochlea, meaning that they are unable to use conventional hearing aids or cochlear implants. The service includes multi-disciplinary assessment, surgical implantation and rehabilitation (including maintenance of the implant). Specialist ear, nose and throat services for children include services provided by specialist ear, nose and throat paediatric surgery centres including outreach when delivered as part of a provider network. The service includes:</p> <ul style="list-style-type: none"> • surgical management of rare conditions • surgical management of more common conditions when the child or the procedure is high risk (including the need for paediatric intensive care or specialist anaesthetic management) • provision of specified procedures • specialist audiology services. <p>1.3 How the service is differentiated from services falling within the responsibilities of other commissioners NHS England will commission this highly specialised service from designated specialised centre(s) who are already commissioned to provide specialist ear, nose and throat paediatric surgery. Children requiring treatment by this service</p>

are regarded as highly complex requiring highly specialist input.

This service is commissioned by NHS England because the number of individuals requiring the service is very small; the cost of providing the service is high because of the specialist interventions and the number of staff trained to provide this service is extremely small.

CCGs do not commission any elements of this service.

Activity is identified via local data flows, which will apply to highly specialised auditory brainstem implant centre(s) only.

2. Care pathway and clinical dependencies

2.1 Care pathway

Children (aged 5 and under) would normally be referred to this highly specialised service by a local auditory implant centre, where, following initial assessment, they are considered unlikely to gain adequate benefit from conventional well-fitted hearing aids or cochlear implants.

Following referral, all children will undergo further comprehensive assessment by a specialist multi-disciplinary team to assess suitability for auditory brainstem implantation.

The multidisciplinary team

The highly specialised auditory brainstem implant team will be embedded within a larger specialised paediatric audiology implant team. The service will be led by an identified member of the core multidisciplinary team (MDT), which will include:

- Healthcare Scientist (Audiology)
- Speech & Language Therapist
- Consultant ENT surgeons who specialise in both cochlear implantation and skull base surgery
- Consultant Neurosurgeons who specialise in skull base surgery
- Administrator.

In addition to the core MDT, the service will have additional named specialists with whom they work:

- Consultant Neuroradiologist
- Paediatric Clinical Psychologist (with experience of working with hearing impaired children and/or complex needs)
- Consultant paediatric neuro-anaesthetist
- Rehabilitation Therapist
- Teacher of the Deaf
- Paediatric Ophthalmologist
- Paediatric Cardiologist
- Paediatric Neurologist

All team members should be suitably qualified and registered with accredited

professional bodies and be highly experienced in their clinical specialty. This experience may have been gained in implanting ABIs in adults who have neurofibromatosis type 2. In addition, all team members will have training in deaf awareness and clinical team members should attend regular training in developments within the field of cochlear and auditory brainstem implantation.

Assessment will include the following:

- Audiological assessments, including otoscopy, age-appropriate behavioural hearing assessment, objective hearing assessments, hearing aid evaluation, aided speech perception testing, balance function testing as required.
- Electrophysiology of the auditory nerve, via electrical auditory brainstem response (eABR) (as required).
- Medical assessment, including clinical history, physical examination, fitness for surgery, radiological evaluation via MRI and CT imaging.
- Rehabilitative team assessment, including speech and language skills, functional listening, quality of life, ability to participate in rehabilitation programme, availability of support & liaison with local services.
- Thorough assessment of child's development and cognitive ability.
- Provision of support to access further appropriate services and care for children with complex and special needs.
- Detailed discussion with parent / carer regarding understanding and expectations of implantation.
- Provision of information about voluntary services and support groups including contact with deaf patients of a similar age (and their families for paediatric patients) who are users of auditory brainstem implants, either face to face or via alternative media.
- Parents/carers will be provided with written information to help them make informed decisions about their child's healthcare at appropriate points within the assessment process. This will include information about potential risks and benefits of surgery.
- Parents/carers will be given appropriate time and space to consider all the information and the implications of implantation, prior to providing informed consent to treatment.

Children not suitable for the device

If, following a multi-disciplinary team (MDT) assessment, it is determined that a child is not suitable for an auditory brainstem implant, the service will ensure that parents/carers and the referrer are provided with the following prior to discharge from the service:

- A clear explanation as to why an ABI is considered to be unsuitable.
- Recommendations for future management, and referral for other equipment and /or services if appropriate.
- An opportunity to discuss the outcome of the assessment with a member of the MDT.
- An opportunity to request an independent second opinion by another commissioned ABI centre in England.

Children suitable for the device

For children who meet the suitability criteria, the service will provide appropriate surgical auditory brainstem implantation. Verbal information should be supported by a written summary to the parents / carers whenever indicated. Throughout the assessment period parents / carers should have a clear understanding of the main benefits and limitations of implantation. They should demonstrate that they have realistic expectations of auditory brainstem implantation.

Device selection

The auditory brainstem implant (ABI) device offered will:

- have a proven track record for safety and reliability
- conform to the recommendations of the Medical and Health Care Products Regulatory Agency (MHRA)
- comply with terms and conditions of the purchasing body
- have high quality clinical and technical support available from the manufacturer
- meet national purchasing requirements.

Surgical implantation

- The operation will be completed by an experienced, specialist team comprising consultant neurosurgeon, ENT surgeon and paediatric neuro-anaesthetist. Implantation must be carried out by appropriately qualified surgeons who have an adequate caseload to maintain surgical skills and optimise outcomes. Anaesthetics must be carried out by appropriately qualified anaesthetists carrying out an adequate caseload of neurosurgery to maintain skills and optimise outcomes.
- Intra-operative testing will be completed by experienced healthcare scientists.
- A minimum of an overnight admission within a paediatric high dependency setting postoperatively is required, with the option for paediatric intensive care if necessary, followed by admission to a paediatric neurosurgical ward until discharge.
- Surgical facilities should afford appropriate NHS standards of safety, accessibility, cleanliness, privacy and dignity and also take into account communication needs of deaf children.

Post-operative care

Prior to discharge the child and parents / carers should receive:

- a medical check of the surgical site and device placement
- written information regarding care of the wound/ear and pain management post operatively
- written guidelines on what to do should medical /surgical problems arise
- advice regarding health and safety with an ABI
- safety guidelines written by the manufacturers (These currently refer to children over the age of 12, but should be provided to all families, explaining that they also apply to younger children until such time as specific advice is available for this age group).
- a date for post-operative / pre-activation medical follow-up.

Device activation and audiological management

Initial activation should be undertaken by an experienced specialist team and should take place following medical advice after the post-operative check-up. For initial activation, children will be admitted, under general anaesthetic, to allow for repeat intraoperative testing; second activation will take place with the child awake and with access to cardiac monitoring, pulse oximetry and resuscitation equipment.

Audiological management will include:

- initial activation and programming of speech processor
- ongoing speech processor programming and assessment dependent on individual need
- frequent audiology outpatient appointments may be required in the first two years following activation. This will reduce to annual review thereafter, or more frequently, depending on the needs of the child.

Rehabilitation

Rehabilitation will be undertaken by an experienced specialist team who will ensure that the rehabilitation needs of the child will be appropriately addressed.

The service must be able to provide all elements of rehabilitation and long term follow up, in accordance with the needs of each child. For prelingually deafened children, it may not be possible by the end of one year post implant, to identify their developmental trajectory and hence their future support needs.

Recognised & validated developmental age appropriate audiological and speech perception measures should be performed on at least two occasions in the first year following surgery. Other rehabilitation requirements may include:

- sound awareness training
- communication skills training
- listening and language skills development training and / or support on an individual child needs basis
- training provided at the centre and as outreach where appropriate
- advice to children and their parents / carers on care and use of the implant
- advice to other organisations supporting the child.

Device maintenance

Parents / carers will be trained by the service to carry out simple trouble-shooting and maintenance such as visual inspection of external parts, including any leads and adaptors, battery replacement and subjective listening checks (where possible).

For more complex maintenance needs, the auditory brainstem implant service will provide advice. The service should aim to resolve repair issues within two working days. Adequate spares / replacements of external equipment must be available as required. Replacement equipment should be issued or despatched on the same or next working day. Speech processor batteries should be available to implant users either from the cochlear implant programme or from a local audiology department by prior agreement.

Matters regarding lost processor(s), upgrades of speech processor(s) and other related issues will be managed in line with the policy for the specialist cochlear implant service.

Device failure

The service will provide re-implantation where possible (under the manufacturers 10 year warranty) in the case of implant failure. If an implant fails after this time, the cost for the new device and associated surgery and programming will be requested from the commissioner. The device failure should be reported on-line to the MHRA Adverse Incidents section. If device failure is suspected the child must be offered an appointment promptly (within seven working days) to check the external and internal components of the implant device.

Data collection

Records of measurement of all programmes installed in the software, and of all tests performed, must be kept on file. Although children will be seen frequently, for the purposes of clinical audit, progress with the ABI should be monitored through at least two assessments in the first year and then at a minimum annually thereafter. These tests should include the child's ability to hear sounds and speech, as well as assessment of quality of life.

All records and measurements should be available on request and provided, with parental consent, to other parties, who may have a legitimate reason for using them, e.g. education, health services etc. The service will be required to engage in clinical audit and service outcome monitoring, sharing pseudonymised data with commissioners and others as appropriate.

2.2 Interdependence with other services

Assessment:

Radiology (*mandatory co-location on same site*)

Paediatric cardiology (*mandatory co-location on same site*)

Paediatric neurology (*mandatory co-location on same site*)

Paediatric clinical psychology (*mandatory co-location on same site*)

Surgery:

Paediatric neurosurgery (*mandatory co-location on same site*)

Paediatric intensive care (*mandatory co-location on same site*)

Neurophysiology (*mandatory co-location on same site*)

Follow-up and rehabilitation:

Radiology assessment (*mandatory co-location on same site*)

Paediatric Audiology (*mandatory co-location on same site*)

Paediatric Speech and Language Therapy (*mandatory co-location on same site*)

Other interactions include:

GPs and Primary Care Practitioners

NHS Audiology Service

NHS New Born Hearing Screening Programme

Educational Services (including Teacher of the Deaf or other specialist teaching services, Social Services including Social Worker for the Deaf, Occupational

Therapy, Physiotherapy, and Educational Psychology).
Voluntary Sector

3. Population covered and population needs

3.1 Population covered by this specification

This service specification relates to children and covers the population defined as the commissioning responsibility of NHS England as set out in “*Who Pays? Determining responsibility for payments to providers*” (2013) guidance <https://www.england.nhs.uk/wp-content/uploads/2014/05/who-pays.pdf> (or subsequent published updates).

Commissioning arrangements for the devolved nations in relation to this service are as set out in “*UK-wide Commissioning Arrangements of Highly Specialised Services*” <https://www.england.nhs.uk/publication/nhs-providers-of-highly-specialised-services/>

3.2 Population needs

Children aged 5 and under with profound deafness, who have no functional hearing as a result of congenital abnormalities affecting the auditory nerve or the cochleae, thus rendering them unable to gain adequate benefit from conventional well-fitted hearing aids or cochlear implants.

It is estimated that about 15 children per annum would be assessed for auditory brainstem implantation and that approximately 6 would go on to have the surgery.

3.3 Expected significant future demographic changes

There are no significant demographic changes expected.

3.4 Evidence base

This specification has been developed on the basis of the evidence review completed during the development of the published Clinical Commissioning Policy: Auditory brainstem implant with congenital abnormalities of the auditory nerves of cochleae: <https://www.england.nhs.uk/wp-content/uploads/2016/12/clin-comm-pol-16062P.pdf>

The published evidence all consists of case series with no randomized controlled trials. In the case of cochlear aplasia, however, it is clear that without treatment the patient will remain without functional hearing.

A systematic review (Merkus 2014) review emphasizes the importance of correct patient selection.

The large series from Colletti (2014) shows that good results can be obtained in many (but not all) patients when selected and treated at an expert centre. On a

standard measure of hearing, 30 out of 64 consecutive children treated with an ABI achieved a score of 4 or better on a 7 point scale of hearing (Categories Auditory Performance CAP), of whom 20 were able to understand free speech (score 5/7). Higher scores were achieved in children treated young, and in those with no other developmental disabilities.

In expert hands, the complication rates are similar to cochlear implantation with a major complication rate of approximately 1% (Colletti 2010). However the severity of complications is greater as these include intracranial complications such as stroke, bleeding and meningitis with the potential for permanent neurological dysfunction.

Consensus statement: Long-term results of ABI in children with complex inner ear malformations and decision making between CI and ABI.

Sennaroğlu L, Colletti V, Lenarz T, Manrique M, Laszig R, Rask-Andersen H, Göksu N, Offeciers E, Saeed S, Behr R, Bayazit Y, Casselman J, Freeman S, Kileny P, Lee DJ, Shannon RV, Kameswaran M, Hagr A, Zarowski A, Schwartz MS, Bilginer B, Kishore A, Sennaroğlu G, Yücel E, Saraç S, Ataş A, Colletti L, O'Driscoll M, Moon IS, Gärtner L, Huarte A, Nyberg G, Mocan BÖ, Atay G, Bajin MD, Çınar BÇ, Batuk MÖ, Yaralı M, Aydınli FE, Aslan F, Kirazlı MC, Özkan HB, Hans JM, Kosaner J, Polak M. Cochlear Implants Int. 2016 Jul;17(4):163-171.

4. Outcomes and applicable quality standards

4.1 Quality statement: Aim of service

The aim of the auditory brainstem implant service is to improve the hearing and quality of life for those with congenital abnormalities of the auditory nerves or the cochlear, rendering them unable to gain adequate benefit from conventional well-fitted hearing aids or cochlear implants.

To promote the normal developmental processes of auditory awareness and spoken language development recognising that this is unlikely to fully occur in the majority of prelingually deafened children.

The service aims to ensure equitable access throughout England to all children who might benefit from an Auditory Brainstem Implant, ensuring that they are assessed in expert centres, with the right children selected for treatment and that children in whom an ABI is inserted receive the necessary device tuning and support for optimal benefit from the device.

NHS outcomes framework domains

Domain 1	Preventing people from dying prematurely	X
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Domain 2	Enhancing quality of life for people with long-term conditions	x
Domain 3	Helping people to recover from episodes of ill-health or following injury	x
Domain 4	Ensuring people have a positive experience of care	x
Domain 5	Treating and caring for people in safe environment and protecting them from avoidable harm	x

4.2 Indicators include:

Number	Indicator	Data Source	Outcome Framework Domain	CQC Key question
Clinical Outcomes				
101	mean waiting time for referral to first appointment	Provider	2,3,5	Effective
102	number of patients return to theatre within 30 days	Provider	2,3,5	Effective
103	30 day mortality	Provider	2,3,5	Effective
104	number of patients with major complications	Provider	2,3,5	Effective
105	number of patients with minor complication	Provider	2,3,5	Effective
106	number of patients failure to implant	Provider	2,3,5	Effective
107	Number of patients with wound infection	Provider	2,3,5	Effective
108	% patients with improved soundfield hearing at 24 months	Provider	2,3,5	Effective
109	% patients with improved CAP score at 24 months	Provider	2,3,5	Effective
110	Mean length of stay in critical care	Provider	2,3,5	Effective
111	Mean length of stay in hospital	Provider	2,3,5	Effective

112	% patients using device	Provider	2,3,5	Effective
Patient Experience				
201	There is information for parents /carers	Self declaration	4	caring, responsive
202	The centre reviews feedback from parents/carers	Self declaration	4	caring, responsive
203	Parents/carers are given training on the use and care of the ABI	Self declaration	4	caring, responsive
204	Parents/carers are allocated a clinical key worker	Self declaration	4	caring, responsive
Structure and Process				
001	The centre is part of a network for cochlear implant services	Self declaration	2,3,5	Effective safe
002	There is a specialist multidisciplinary team	Self declaration	2,3,5	Effective safe
003	Staff receive training specific to ABI	Self declaration	2,3,5	Effective safe
004	There are BSL and interpreters	Self declaration	4	caring, responsive
005	Patients have a multidisciplinary assessment	Self declaration	2,3,5	Effective safe
006	There is a device management policy	Self declaration	2,3,5	Effective safe
007	There are clinical guidelines	Self declaration	2,3,5	Effective safe
008	There are patient pathways in place	Self declaration	2,3,5	Effective safe

Detailed definitions of indicators setting out how they will be measured are included in schedule 6.

4.3 Commissioned providers are required to participate in annual quality assurance and collect and submit data to support the assessment of compliance with the service specification as set out in Schedule 4A-C. Commissioned providers will be required to work collaboratively; meeting at least annually, to share clinical outcomes, adverse events, discuss relevant published literature and developments in the field to ensure the safe and effective provision of an equitable national service that meets the needs of the identified population.

4.4 Applicable CQUIN goals are set out in Schedule 4D

5. Applicable service standards

5.1 Applicable obligatory national standards

There are no obligatory national standards available for this service. (See consensus statement above)

5.2 Other applicable national standards to be met by commissioned providers

The services must be provided in specialist cochlear implant centres which may be hospital or university based. As the rehabilitation required to support successful use of auditory brainstem implants can be intense, these services would be expected to be provided by the nearest ABI centre or on an outreach/shared care basis with local cochlear implant services where appropriate.

The service must have access to appropriately calibrated and up-to-date equipment and facilities to enable all appropriate assessments to be undertaken. Audiological testing will be performed in appropriately sound treated rooms where possible such that the ambient noise levels are compliant with the BBS EN ISO 8253-1:1998 standard, Acoustics: - Audiometric test Methods – part 1.

The service must have facilities appropriate NHS standards of safety, accessibility, cleanliness, privacy and dignity and also take into account communication needs of deaf patients and requirements for specific assessments such as sound-proofing. The design and layout must take into account the needs of families and young children within their client group. Services, facilities and accommodation must comply with current British standards and the Equality Act 2010. All facilities must comply with Health and Safety Executive regulations.

5.3 Other applicable local standards

Given the very high level of expertise required to deliver the service, the successful provider(s) will be required to seek international mentorship from a clinician who is a recognised leader in this field.

6. Designated providers (if applicable)

To be confirmed following provider selection process.

7. Abbreviation and acronyms explained

The following abbreviations and acronyms have been used in this document:

ABI - Auditory Brainstem Implant
BCIG – British Cochlear Implant Group
BSL - British Sign Language
CCG - Clinical Commissioning Group
CRG - Clinical Reference Group
CT Scan - Computed Tomography Scan
eABR - Electrical Auditory Brainstem Response
ENT - Ear, Nose and Throat
MDT - Multidisciplinary Team
MHRA - Medicines and Healthcare products Regulatory Agency
MRI - Magnetic Resonance Imaging

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