Specialised Services Circular

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Cochlear Implantation and patients with ANSD

Circulation

For action	**For information**
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Background

NHS England took over the responsibility for commissioning Cochlear Implantation in April 2013. A Commissioning Policy was developed and agreed following public consultation which clearly states that CI would be commissioned in line NICE TAG 166, which was issued in January 2009.

A hearing condition currently identified as ANSD [auditory neuropathy spectrum disorder] has always produced assessments that can sit across different domains of the NICE TAG. The purpose of this document is to clarify this condition with respect to the NICE TAG and commissioning. The condition is more commonly seen in

children and this circular is aimed at clarification especially for children.

The NICE TAG 166 states that cochlear implantation is recommended as an option for people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.

These two parameters are then further clarified as:

- Severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz without acoustic hearing aids
- Adequate benefit from acoustic hearing aids is defined for this guidance as:

  - for adults, a score of 50% or greater on Bamford–Kowal–Bench (BKB) sentence testing at a sound intensity of 70 dB SPL
  - for children, speech, language and listening skills appropriate to age, developmental stage and cognitive ability.

An individual with ANSD MAY have thresholds as measured at individual frequencies that appear “better” than those defined above but whose functional hearing performance is consistent with profound hearing loss and whose speech, language and listening skills are significantly below expected levels. As such the Specialised Ear Surgery Clinical Reference Group (CRG) has always been in agreement that this group of patients fall within the scope of NICE TAG 166 and NHS England’s Clinical Commissioning Policy and therefore a cochlear implant would be routinely commissioned.

The CRG has however recently become aware that some Area Teams view is that patients with ANSD are not routinely funded and patients are subsequently being turned down at IFR panels. AS such the CRG has been asked to produce this circular to clarify how this group of patients meet the criteria and to provide any additional advice for Area Teams and Providers.

Summary

The underlying cause of ANSD is poorly understood and it is recognised that there may be some spontaneous improvement with age. It is most frequently noted in premature babies.

Due to the difficulty in diagnosing this group of children and in order to provide assurance that Providers are treating patients who fit within the existing clinical criteria the CRG fully supports the recent protocol issued by the British Cochlear Implant Group as outlined below:

1. The diagnosis of ANSD and testing used must be clearly specified and conform to the recommendations in the guidelines [enclosed]

2. All children must have clear evidence of continued monitoring and

assessment to demonstrate the lack of improvement with maturity. The exact tests and assessments cannot be individually specified but the most recent should be conducted after the child has passed 15 months corrected age.

3. All children must have been subject to a hearing aid trial of at least 3 months duration in association with testing outlined above to demonstrate evidence of progress [or lack of it] in order to demonstrate that they meet the NICE TAG 166 criteria.

In order to confirm diagnosis of ANSD the following tests should be used:

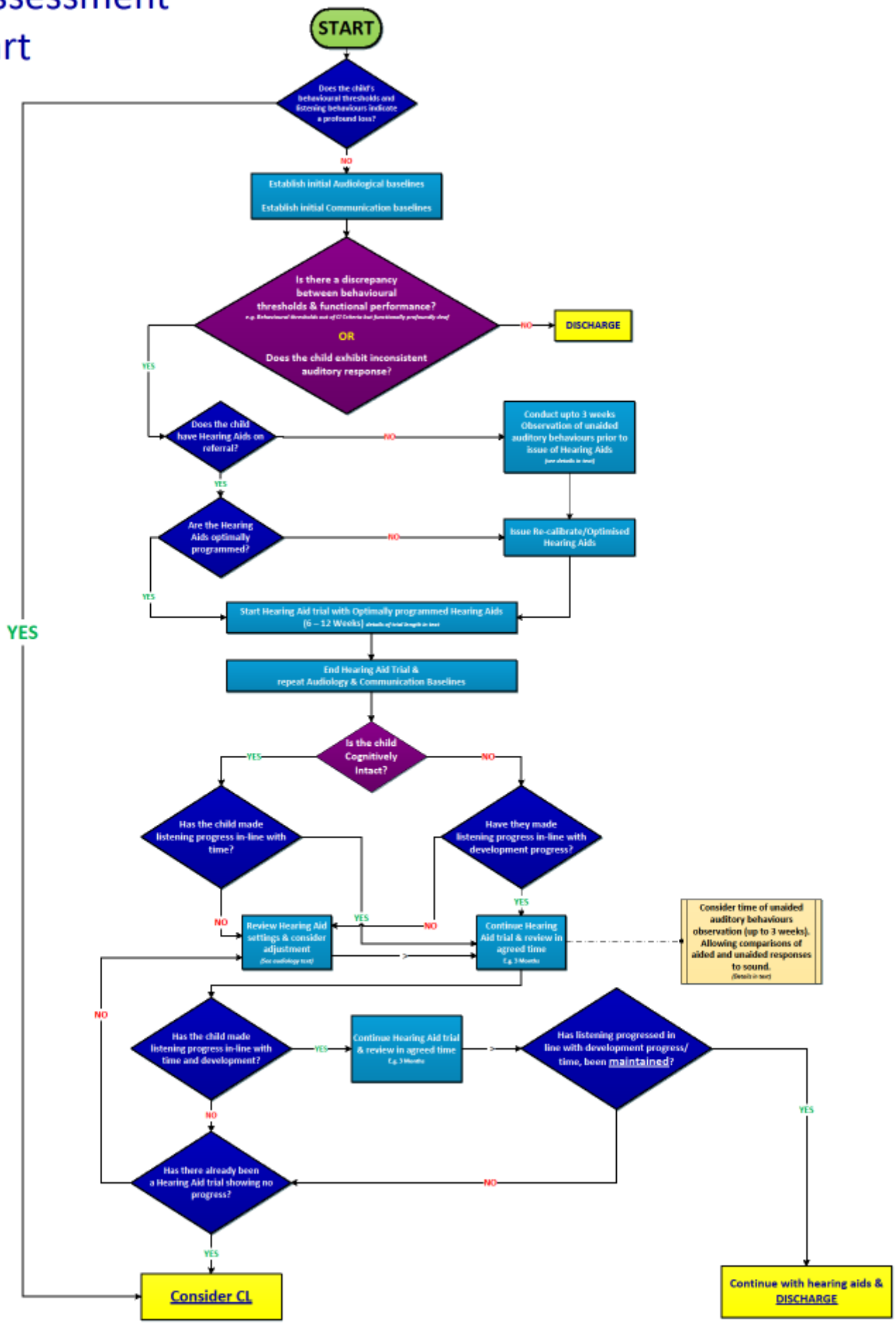
- Acoustic reflexes, when possible. Acoustic reflexes are likely to be absent or elevated in cases of ANSD (Berlin, Hood et al. 2005)
- Otoacoustic emissions
- Repeat ABR test at minimum 9 months corrected age, including cochlear microphonic testing according to NHSP Guidelines for Cochlear Microphonic Testing (Lightfoot, Stevens et al. 2011). It is possible that maturation of the ABR response can be delayed by up to 18 months or more. Literature on this subject reports cases of recovery of the ABR up to 6-24 months after initial diagnosis (Psarommatis, Riga et al. 2006; Attias and Raveh 2007). There is no evidence to suggest that the maturation effect occurs suddenly after an extended period of no change. Where the result of the repeated ABR shows no evidence of maturation and there is no change in behavioural responses, management should continue as described in the ANSD assessment pathway. Where the result of the repeated ABR shows signs of recovery of the neural response, a different management pathway is required, in which ABR testing is monitored regularly until there is a stable ABR response. This to be performed in parallel with the hearing aid trial.

Action

Area Teams to note that children with ANSD should have access to Cochlear Implantation in line with NHS England's Clinical Commissioning Policy as long as the above criteria is met. Providers must ensure that a child with ANSD has been appropriately assessed and had the opportunity to mature and improve with conventional hearing aids. If this has been done and there is no improvement then cochlear implantation is an appropriate intervention and is consistent with the current NICE TAG 166.

Further Information

ANSD Assessment Flowchart

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