

Multidisciplinary Macroglossia Service for children with Beckwith-Wiedemann Syndrome

This is a national specialised service for children who have macroglossia associated with Beckwith Wiedemann Syndrome (BWS macroglossia). The service was designated in April 2012 by the NHS Advisory Group for National Specialised Services (AGNSS) and is now commissioned as a highly specialised service by NHS England. It is located at Great Ormond Street Hospital for Children NHS Foundation Trust (GOSH).

The GOSH service

Between 1994 and June 2018, 280 children with BWS macroglossia have been followed longitudinally throughout childhood at GOSH. Outcome data has been systematically collected across a range of dimensions both pre and post tongue reduction surgery as well as for children who have not undergone surgery. The GOSH service is therefore embedded in multi-professional evidence based practice. Recommendations are based upon the specialist knowledge of the condition and are consequently specific and time efficient for patients and local professionals. There is no other centre within the UK seeing a large volume of patients with BWS macroglossia and providing long term coordinated care.

Aim of the service

The aim of the service is to prevent or resolve the problems associated with macroglossia in BWS in a timely way and to provide best current practice. The problems caused by the macroglossia include: difficulties with feeding and speech, the position of the teeth and lower jaw, increased drooling and altered facial appearance. In some cases respiratory difficulties can occur.

Service provision

- Assessment of the impact of the macroglossia. This includes assessment of feeding, speech, drooling, oral motor skills, malocclusion and facial growth.
- Differential diagnosis between functional difficulties caused by the macroglossia and other developmental/medical problems.
- Specialist management advice for the functional difficulties caused by the macroglossia.
- Information on BWS macroglossia, the long term outcomes for children with the condition and the management options.
- MDT decision regarding the appropriateness and timing of tongue reduction surgery.
- Tongue reduction surgery carried out by the specialist surgeon. Note: not all children will require surgery.
- Preparation for surgery and advice/management in the post-operative recovery period.
- Liaison with local services
- Follow up as appropriate.

Referrals to the service

The child must have a diagnosis of macroglossia associated with BWS. A written referral stating the means of diagnosis of Beckwith-Wiedemann syndrome is required, enclosing a copy of the molecular genetics results. Where molecular confirmation has not been possible (as is currently the case in approximately 20%) the child should fulfill clinical diagnostic criteria for BWS. The child must be 6 months at age of referral so that the macroglossia is stable. Babies below 6 months of age who have feeding difficulties should be referred to their local children's speech and language therapy dysphagia service.

Core clinicians in the service

Caroleen Shipster

Specialist Speech & Language Therapist and Lead Clinician for the Beckwith-Wiedemann with Macroglossia Service

Professor David Dunaway CBE

Consultant Craniofacial and Plastic Surgeon

Mr Juling Ong

Consultant Craniofacial and Plastic Surgeon

Mr Robert Evans

Consultant Orthodontist

Nivi Behari

Specialist Speech & Language Therapist
Gaynor Osman and Brigitte Harrison
Clinical Nurse Specialists

Referrals to the service should be sent to:

Caroleen Shipster, Lead Clinician for the BWS Macroglossia Service

c/o The BWS Macroglossia Service Co-ordinator, Claire Yule, Speech and Language Therapy Department,
Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH, UK.

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