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SURVEILLANCE OF SEVERE MICROCEPHALY IN THE UK & IRELAND (SSM-UKI)

Abstract

Microcephaly describes a baby with a 'small head', who has experienced poor head growth before or after birth. Severe microcephaly is defined by the World Health Organization (WHO) as a head circumference more than 3 standard deviations (<-3SD) below the mean for age and sex. This rare condition may be associated with abnormal brain structure or neurodevelopment, and with disability, although some babies will develop normally. Many different causes of microcephaly have been described, including genetic disorders, exposure during pregnancy to environmental toxins, certain drugs, infection or malnutrition. Two important infections in the mother that may cause microcephaly are rubella (German measles) and Zika virus.

This study will find out how many UK babies are born with microcephaly each year and how seriously this affects their health and developmental outcomes, including problems with hearing and vision. It will provide a better picture of the care and support these babies and their families currently receive. This will allow us to make sure the right services are available to meet the future needs of children and families. Importantly, through comparison with congenital anomaly registers, it will also provide us with a way to monitor future changes in the frequency of microcephaly.

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Background

Measurement of head circumference is part of the routine newborn examination, however microcephaly is often not recognised until later in infancy or childhood as a consequence of abnormal neurodevelopment. Although congenital anomaly registers suggest that microcephaly is uncommon in the UK, this system relies on passive reporting so may underestimate the actual numbers affected.

When microcephaly is associated with brain abnormalities, then babies may have a wide range of serious neurodevelopmental and cognitive problems, including epilepsy, cerebral palsy, learning disabilities, hearing loss or vision problems. Although research into the longer-term outcomes of children with severe microcephaly is limited, in one case series around half of affected children had an IQ below 70 at seven years of age. Most children with severe microcephaly will therefore experience a significant lifelong impact on their neurodevelopment, education, social and emotional wellbeing.

Timely investigation, confirmation of neurodisability, genetic counselling and access to services are essential to support families with a child affected by microcephaly. Multi-disciplinary clinical management is focused at mitigating neurodevelopmental problems and learning difficulties. In the UK, insufficient information to inform care is available from previous studies or existing 'routine' data sources. Findings from this study will therefore inform the development of clinical guidelines for microcephaly and provide robust evidence to support the future development of high quality clinical services to improve outcomes.

Coverage

United Kingdom and the Republic of Ireland

Duration

October 2017 – October 2018 (13 months) with follow-up for two years

PTO

Research Questions

1. Determine the incidence of severe microcephaly in children aged up to 12 months old in the UK and Ireland
2. To report clinical features at presentation, including associated comorbidities, congenital anomalies and neurological features, vision and hearing impairment
3. To describe variations in clinical investigation and management
4. To describe clinical outcomes at age one and two years, including neurodisability, vision and hearing problems, and use of health and social services
5. To compare the number of cases reported through the BPSU and routine passive reporting systems, such as congenital anomaly registers.

Case definition

Any live born infant up to and including 12 months of age who has a head circumference more than 3 standard deviations below the mean (<-3SD), adjusted for age-, sex- and gestation (using the UK-WHO growth chart standard or UK90 growth chart for babies under 2 weeks of age).

Excluding: infants with anencephaly, and/or similar neural tube defects, (where the standard measurement of occipito-frontal circumference is not appropriate).

Reporting instructions

Please report any live born child aged up to and including 12 months of age who:

- has been diagnosed with microcephaly in the past month,

and/or noted to have a head circumference that is:

- more than three standard deviations below the mean (<-3SD) for gestational age and sex, **or** below the <0.4th percentile for age and sex.

Use your standard growth chart.

If you are unsure whether the child meets the case definition, for example an infant has only had one abnormal head measurement, please just report the child or contact us to the principal investigator.

Excluding: Please do not report babies with anencephaly.

Methods

1. Active surveillance through the BPSU for 13 months with two years follow-up of outcomes.
2. Data will be collected from reporting clinicians at initial notification (and 12 and 24 months later) using standardised online questionnaires for data collection.
3. Information will be collected about socio-demographic variables, growth and neurodevelopment, and associated comorbidities.
4. The follow up questionnaire will confirm neurodevelopmental outcomes and disability, wider health outcomes, and service use.

Ethics approval

The study has been approved by East of Scotland Research Ethics Committee (reference: 17-ES-088) HRA Confidentiality Advisory Group (reference: 17-CAG-0126) and the Scottish Public Benefit and Privacy Panel (reference: 1718-0184).

Support groups

Sense (www.sense.org.uk)- a national service-providing organisation for children and adults with sensory impairments; **Contact** (www.contact.org.uk) - a charity supporting families of children living with a disability.

Funding

The study is funded by Great Ormond Street Hospital Children's Charity.

References

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