



Congenital Microcephaly

Background

Congenital microcephaly is an abnormal smallness of the head, a congenital condition associated with incomplete brain development. This condition most commonly occurs between the third and fourth month of gestation. Microcephaly is a neural proliferation defect. In the United States, it has been estimated that the incidence of microcephaly is approximately 2–12 babies per 10,000 live births (Centers for Disease Control [CDC], 2018).

Definition

Microcephaly is a rare condition in which the frontal occipital circumference (FOC) is equal to or less than two standard deviations below the mean for age and gender or less than the 10th percentile for gestational age.

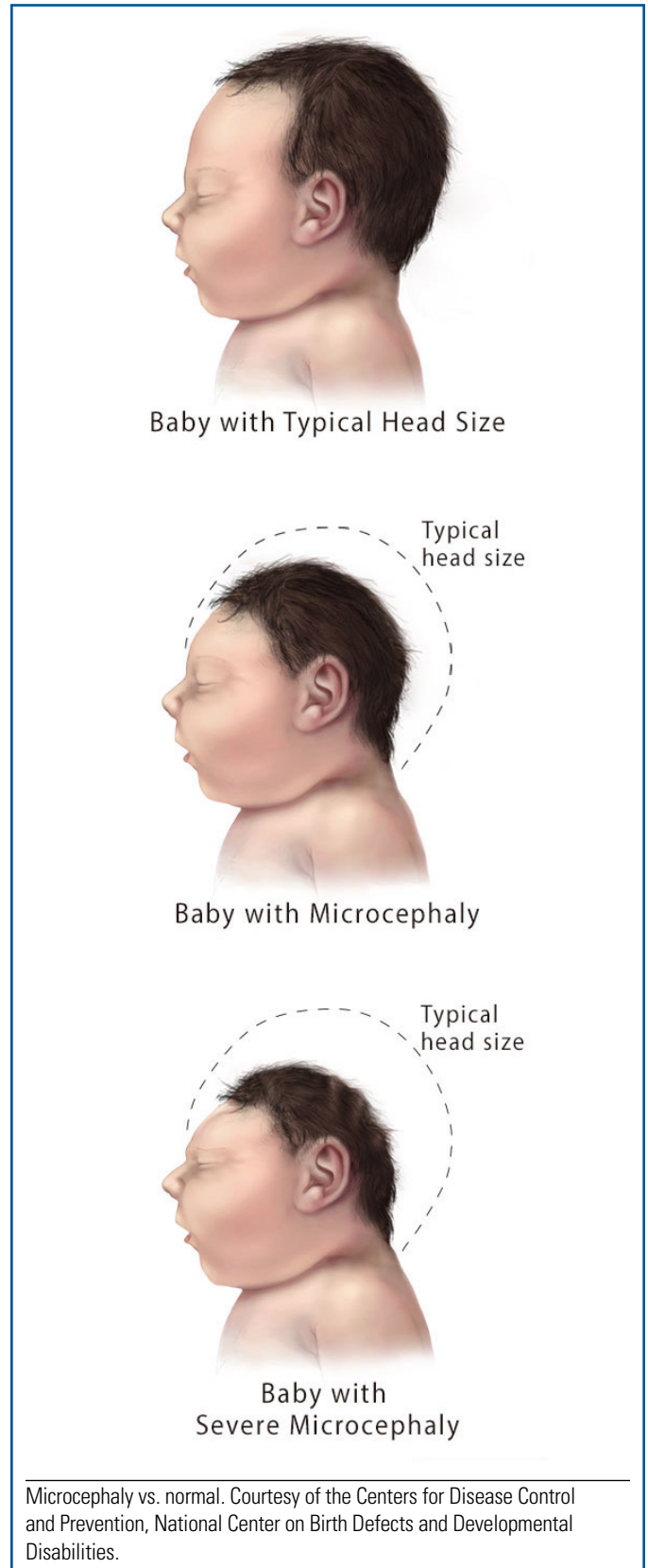
Risk Factors

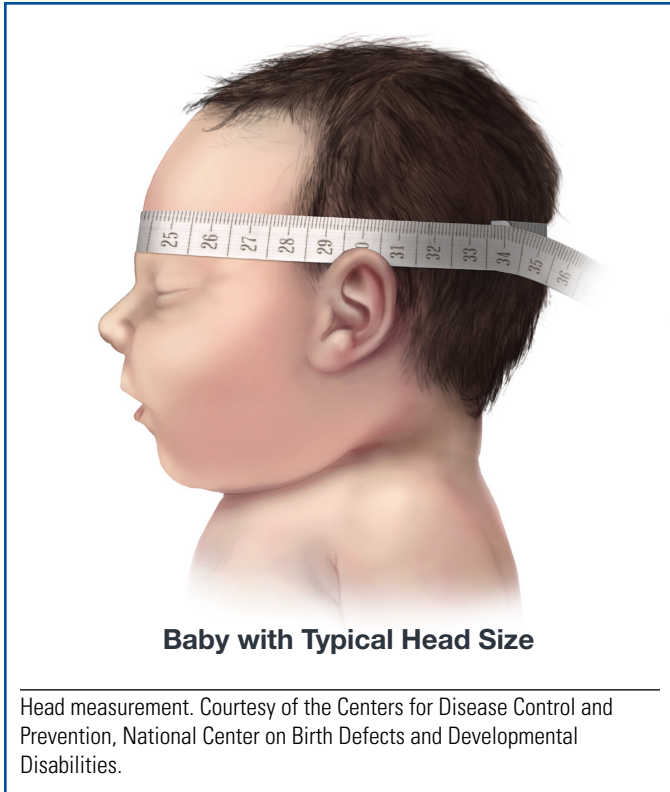
The exact etiology for microcephaly is unknown, but it can be caused by maternal, fetal, or neonatal factors. Maternal causes may include viral infections (toxoplasmosis, syphilis, rubella, cytomegalovirus, or herpes simplex), radiation exposure, metabolic disorders (PKU), exposure to harmful toxins (toxic chemicals, alcohol, and drugs) particularly during the first trimester, genetic conditions, and malnutrition. Fetal causes may include a prenatal or perinatal insult such as inflammation, hypoxia, or an interruption of the blood flow to the brain during a critical period of development, or birth trauma. Malnutrition, hypoxic ischemic encephalopathy, and very low birth weight (less than 1,500 grams at birth) can be neonatal causes. In recent years, maternal infection with the Zika virus during pregnancy has been identified as a risk factor for congenital microcephaly or other severe brain and nervous system disorders.

Associated Problems

Depending on the severity, microcephaly may be associated with other lifelong problems such as

- seizures
- developmental delays, such as problems with speech
- intellectual deficits
- difficulties with movement and balance





- feeding problems, such as difficulty with swallowing
- hearing loss
- visual disturbances.

Clinical Presentation

On visual inspection, the head appears small in relation to the rest of the body. This suspicion is confirmed when the head circumference is measured. Optimally, this measurement is obtained within the first 24 hours of life. Head circumference is measured with a tape that cannot be stretched. Securely wrap the tape around the widest point of the head's circumference to include the broadest part of the forehead above the eyebrows, above the ears, and the most prominent part of the occiput (Tappero & Honeyfield, 2015).

Congenital Zika Syndrome

Infants whose mother's were infected with the Zika virus during pregnancy will need to be closely evaluated for congenital Zika syndrome. This syndrome has five unique features:

- severe microcephaly in which the skull has partially collapsed

- decreased brain tissue with a specific pattern of brain damage
- damage to the back of the eye
- congenital contractures (CDC, 2018)
- hypertonia that restricts body movement after birth.

This infection has also been linked to other brain abnormalities such as brain atrophy, abnormally formed or absent brain structures, hydrocephalus, and neuronal migration disorders.

Diagnosis

Congenital microcephaly may be diagnosed prenatally during ultrasound evaluations, most accurately if the ultrasound is obtained late in the second trimester or early in the third trimester. This condition is confirmed or diagnosed after delivery during physical examination. Further studies may include a computed tomography (CT) or magnetic resonance imaging (MRI) of the head after birth.

Laboratory Findings

Depending on the specific risk factors, additional laboratory tests may be ordered. For example, if the cause is suspected to be a viral infection, labs may be drawn to confirm or rule out this cause.

Treatment

Microcephaly is a lifelong condition in which there are no treatment options available. Because the effects of microcephaly can range from mild to severe, care should be tailored to the individual child. Early intervention programs that include speech, physical, and occupational therapies will maximize the potential for these children.

References

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Bibliography

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Congenital Microcephaly: Information for Parents

Congenital microcephaly is a rare condition in which the baby's head does not grow as expected in utero. During pregnancy, the fetus's head increases in size because his or her brain is growing. Therefore, when a baby has a smaller head than expected, the concern is that the brain is not growing as it should. This condition most commonly occurs during the first trimester of the pregnancy.

Microcephaly also may develop after birth (acquired microcephaly) and would be identified during well-baby checkups. In the United States, about five of every 10,000 babies born will have microcephaly.

The exact reason for microcephaly is not always clear, but it can be caused by many different things. Microcephaly may be the result of a viral infection the mother had during pregnancy (especially during the first few months of pregnancy). Other causes may include radiation exposure; metabolic disorders such as maternal phenylketonuria (PKU) disease; exposure to toxic chemicals, alcohol, or drugs (especially during the first trimester); genetic conditions; and decreased nutrition during pregnancy. Other causes may include inflammation, an interruption of the blood flow and oxygen to the brain of the baby during the critical period of development, and birth trauma. In recent years, research has shown that babies of mothers who were infected with the Zika virus during pregnancy are at risk for developing microcephaly or other severe brain and nervous system disorders.

Microcephaly can be mild or severe. If severe, microcephaly may be associated with other lifelong problems such as

- seizures
- developmental delays, such as speech problems
- intellectual deficits, such as learning difficulties
- difficulties with movement and balance
- feeding problems, such as difficulty with swallowing
- hearing loss
- visual issues.

When looking at your baby, his or her head appears small in proportion to the rest of the body. This suspicion is confirmed when the head circumference is measured. Optimally, this measurement is obtained soon after birth. On some babies with microcephaly, you may also notice a backward slope of the forehead.

Congenital microcephaly may be diagnosed before birth during ultrasound evaluations, especially when the ultrasound is done late in the second trimester or early in the third trimester. Sometimes, microcephaly is diagnosed after delivery, when the physician or nurse practitioner does their physical exam. Your baby may need a scan (CT or MRI) of the head to further evaluate this issue.

Depending on the risk factors for your baby, additional tests may be ordered. For example, if the suspected cause is a viral infection, blood will be drawn to confirm or rule this out.

Microcephaly is a lifelong condition, and currently, there are no treatments for it. Because the effects of microcephaly can range from mild to severe, each baby will have a different care plan. Early intervention programs that include speech, physical, and occupational therapies will be the most helpful for these children.

Babies infected with the Zika virus before birth will need to be closely evaluated for congenital Zika syndrome. This syndrome has five unique features:

- severe microcephaly
- decreased brain tissue with a specific pattern of brain damage
- damage to the back of the eye
- congenital contractures (a condition that shortens and/or tightens the muscles and tendons in the arms and legs, leading to deformity)
- increased muscle tone that restricts body movement after birth.



If your baby has had this infection, he or she also may have other brain abnormalities. Because of the lifelong consequences of microcephaly, it is very important that your baby has regular well-baby visits with his or her primary care or pediatric providers.