[Template:Redirect](/wiki/Template:Redirect" \o "Template:Redirect) [Template:Infobox medical condition](/wiki/Template:Infobox_medical_condition) **Microcephaly** is a medical condition in which the [brain](/wiki/Brain) does not develop properly resulting in a smaller than normal [head](/wiki/Head).<ref name=NIH2015>[Template:Cite web](/wiki/Template:Cite_web)</ref> Microcephaly may be [present at birth](/wiki/Congenital) or it may develop in the first few years of life.<ref name=NIH2015/> Often people with the disorder have an [intellectual disability](/wiki/Intellectual_disability), poor motor function, poor speech, abnormal facial features, [seizures](/wiki/Seizures), and [dwarfism](/wiki/Dwarfism).<ref name=NIH2015/>

The disorder may stem from a wide variety of conditions that cause abnormal growth of the brain, or from [syndromes](/wiki/Syndrome) associated with [chromosomal](/wiki/Chromosome) abnormalities. A [homozygous](/wiki/Homozygous#Homozygous) [mutation](/wiki/Mutation) in one of the [*microcephalin*](/wiki/Microcephalin) genes causes primary microcephaly.[[1]](#cite_note-1)[[2]](#cite_note-2) It serves as an important [neurological](/wiki/Neurological) indication or warning sign, but no uniformity exists in its definition. It is usually defined as a [head circumference](/wiki/Human_head#Anthropometry) (HC) more than two [standard deviations](/wiki/Standard_deviation) below the mean for age and sex.[[3]](#cite_note-3)[[4]](#cite_note-4) Some academics advocate defining it as head circumference more than three standard deviations below the mean for the age and sex.[[5]](#cite_note-5) There is no specific treatment that returns the head size to normal.<ref name=NIH2015/> In general, life expectancy for individuals with microcephaly is reduced and the [prognosis](/wiki/Prognosis) for normal brain function is poor. Occasionally some will grow normally and develop normal intelligence.<ref name=NIH2015/>

## Contents

* 1 Signs and symptoms[[edit](/index.php?title=(none)&action=edit&section=1)]
* 2 Causes[[edit](/index.php?title=(none)&action=edit&section=2)]
  + 2.1 Congenital[[edit](/index.php?title=(none)&action=edit&section=3)]
  + 2.2 Postnatal onset[[edit](/index.php?title=(none)&action=edit&section=4)]
  + 2.3 Microencephaly[[edit](/index.php?title=(none)&action=edit&section=5)]
  + 2.4 Other[[edit](/index.php?title=(none)&action=edit&section=6)]
* 3 Treatment[[edit](/index.php?title=(none)&action=edit&section=7)]
* 4 History[[edit](/index.php?title=(none)&action=edit&section=8)]
* 5 Notable cases[[edit](/index.php?title=(none)&action=edit&section=9)]
* 6 See also[[edit](/index.php?title=(none)&action=edit&section=10)]
* 7 References[[edit](/index.php?title=(none)&action=edit&section=11)]
* 8 External links[[edit](/index.php?title=(none)&action=edit&section=12)]

## Signs and symptoms[[edit](/index.php?title=(none)&action=edit&section=1)]

Affected newborns generally have striking [neurological](/wiki/Neurological) defects and [seizures](/wiki/Seizure). Severely impaired [intellectual](/wiki/Wikt:intellectual) development is common, but disturbances in [motor functions](/wiki/Motor_function) may not appear until later in life.[Template:Citation needed](/wiki/Template:Citation_needed)

[Infants](/wiki/Infant) with microcephaly are born with either a normal or reduced head size. Subsequently, the head fails to grow, while the face continues to develop at a normal rate, producing a child with a small head and a receding forehead, and a loose, often wrinkled [scalp](/wiki/Scalp). As the child grows older, the smallness of the [skull](/wiki/Human_skull) becomes more obvious, although the entire body also is often underweight and [dwarfed](/wiki/Dwarfism). Development of motor functions and [speech](/wiki/Speech_communication) may be delayed. [Hyperactivity](/wiki/Hyperactive) and intellectual disability are common occurrences, although the degree of each varies. [Convulsions](/wiki/Convulsion) may also occur. Motor ability varies, ranging from [clumsiness](/wiki/Wikt:clumsy) in some to [spastic](/wiki/Spastic) [quadriplegia](/wiki/Quadriplegia) in others.[Template:Citation needed](/wiki/Template:Citation_needed)

## Causes[[edit](/index.php?title=(none)&action=edit&section=2)]

[thumb|Neural scans of a normal-sized skull (left) and a case of microcephaly (right)](/wiki/Image:Microcephaly.png) Microcephaly is a type of [cephalic disorder](/wiki/Cephalic_disorder). It has been classified in two types based on the onset:[[6]](#cite_note-6)

### Congenital[[edit](/index.php?title=(none)&action=edit&section=3)]

[Template:Refimprove section](/wiki/Template:Refimprove_section)

Isolated

1. Familial ([autosomal recessive](/wiki/Autosomal_recessive)) microcephaly
2. [Autosomal dominant](/wiki/Autosomal_dominant) microcephaly
3. [X-linked](/wiki/X-linked) microcephaly
4. Chromosomal (balanced rearrangements and ring chromosome)

Syndromes

* Chromosomal

1. [Poland syndrome](/wiki/Poland_syndrome)
2. [Down syndrome](/wiki/Down_syndrome)
3. [Edward syndrome](/wiki/Edward_syndrome)
4. [Patau syndrome](/wiki/Patau_syndrome)
5. Unbalanced rearrangements

* Contiguous gene deletion

1. 4p deletion ([Wolf–Hirschhorn syndrome](/wiki/Wolf–Hirschhorn_syndrome))
2. 5p deletion ([Cri-du-chat](/wiki/Cri-du-chat))
3. 7q11.23 deletion ([Williams syndrome](/wiki/Williams_syndrome))
4. 22q11 deletion ([DiGeorge syndrome](/wiki/DiGeorge_syndrome))

* Single gene defects

1. [Smith–Lemli–Opitz syndrome](/wiki/Smith–Lemli–Opitz_syndrome)
2. [Seckel syndrome](/wiki/Seckel_syndrome)
3. [Cornelia de Lange syndrome](/wiki/Cornelia_de_Lange_syndrome)
4. [Holoprosencephaly](/wiki/Holoprosencephaly)
5. [Primary microcephaly 4](/wiki/Primary_microcephaly_4)<ref name=Szczepanski2015>Szczepanski S, Hussain MS Sur I, Altmüller J, Thiele H, Abdullah U, Waseem SS, Moawia A, Nürnberg G, Noegel AA, Baig SM, Nürnberg P (2015) A novel homozygous splicing mutation of CASC5 causes primary microcephaly in a large Pakistani family. Hum Genet</ref>

Acquired

* Disruptive injuries

1. [Ischemic stroke](/wiki/Ischemic_stroke)
2. [Hemorrhagic stroke](/wiki/Hemorrhagic_stroke)
3. Death of a monozygotic twin

* [Vertically transmitted infections](/wiki/Vertically_transmitted_infection)

1. [Congenital cytomegalovirus infection](/wiki/Congenital_cytomegalovirus_infection)
2. [Toxoplasmosis](/wiki/Toxoplasmosis)
3. [Congenital rubella syndrome](/wiki/Congenital_rubella_syndrome)
4. [Zika virus](/wiki/Zika_virus) [[7]](#cite_note-7)[[8]](#cite_note-8)[[9]](#cite_note-9)

* Drugs

1. [Fetal hydantoin syndrome](/wiki/Fetal_hydantoin_syndrome)
2. [Fetal alcohol syndrome](/wiki/Fetal_alcohol_syndrome)

Other

1. [Radiation](/wiki/Radiation) exposure to mother
2. Maternal [malnutrition](/wiki/Malnutrition)
3. Maternal [phenylketonuria](/wiki/Phenylketonuria)
4. Poorly controlled [gestational diabetes](/wiki/Gestational_diabetes)
5. [Hyperthermia](/wiki/Hyperthermia)
6. Maternal [hypothyroidism](/wiki/Hypothyroidism)
7. [Placental insufficiency](/wiki/Placental_insufficiency)

### Postnatal onset[[edit](/index.php?title=(none)&action=edit&section=4)]

[Template:Refimprove section](/wiki/Template:Refimprove_section)

Genetic

* Inborn errors of metabolism

1. [Congenital disorder of glycosylation](/wiki/Congenital_disorder_of_glycosylation)
2. [Mitochondrial disorders](/wiki/Mitochondrial_disorders)
3. [Peroxisomal disorder](/wiki/Peroxisomal_disorder)
4. [Glucose transporter](/wiki/Glucose_transporter) defect
5. [Menkes disease](/wiki/Menkes_disease)
6. [Congenital disorders of amino acid metabolism](/wiki/Congenital_disorders_of_amino_acid_metabolism)
7. [Organic acidemia](/wiki/Organic_acidemia)

Syndromes

* Contiguous gene deletion

1. 17p13.3 deletion ([Miller–Dieker syndrome](/wiki/Miller–Dieker_syndrome))

* Single gene defects

1. [Rett syndrome](/wiki/Rett_syndrome) (primarily girls)
2. [Nijmegen breakage syndrome](/wiki/Nijmegen_breakage_syndrome)
3. X-linked [lissencephaly](/wiki/Lissencephaly) with abnormal genitalia
4. [Aicardi–Goutières syndrome](/wiki/Aicardi–Goutières_syndrome)
5. [Ataxia telangiectasia](/wiki/Ataxia_telangiectasia)
6. [Cohen syndrome](/wiki/Cohen_syndrome)
7. [Cockayne syndrome](/wiki/Cockayne_syndrome)

Acquired

* Disruptive injuries

1. [Traumatic brain injury](/wiki/Traumatic_brain_injury)
2. [Hypoxic-ischemic encephalopathy](/wiki/Hypoxic-ischemic_encephalopathy)
3. [Ischemic stroke](/wiki/Ischemic_stroke)
4. [Hemorrhagic stroke](/wiki/Hemorrhagic_stroke)

* Infections

1. Congenital [HIV encephalopathy](/wiki/HIV_encephalopathy)
2. [Meningitis](/wiki/Meningitis)
3. [Encephalitis](/wiki/Encephalitis)

* Toxins

1. [Lead poisoning](/wiki/Lead_poisoning)
2. [Chronic renal failure](/wiki/Chronic_renal_failure)

* Deprivation

1. [Hypothyroidism](/wiki/Hypothyroidism)
2. [Anemia](/wiki/Anemia)
3. [Congenital heart disease](/wiki/Congenital_heart_disease)
4. [Malnutrition](/wiki/Malnutrition)

A genetic factor may play a role in causing some cases of microcephaly. Relationships have been found between [autism](/wiki/Autism), duplications of chromosomes, and [macrocephaly](/wiki/Macrocephaly) on one side. On the other side, a relationship has been found between [schizophrenia](/wiki/Schizophrenia), deletions of chromosomes, and microcephaly.[[10]](#cite_note-10)[[11]](#cite_note-11)[[12]](#cite_note-12) Moreover, an association has been established between common genetic variants within known microcephaly genes (*MCPH1, CDK5RAP2*) and normal variation in brain structure as measured with [magnetic resonance imaging](/wiki/Magnetic_resonance_imaging) (MRI)[Template:Nsmdnsi](/wiki/Template:Nsmdns).e., primarily brain cortical surface area and total brain volume.[[13]](#cite_note-13) The spread of [Aedes](/wiki/Aedes) [mosquito](/wiki/Mosquito)-borne [Zika virus](/wiki/Zika_virus) has been implicated in increasing levels of congenital microcephaly by the [International Society for Infectious Diseases](/wiki/International_Society_for_Infectious_Diseases) and the US [Centers for Disease Control and Prevention](/wiki/Centers_for_Disease_Control_and_Prevention).[[14]](#cite_note-14) Zika can spread from a pregnant woman to her fetus. This can result in other severe brain malformations and birth defects.<ref name=NEJM201604>[Template:Cite journal](/wiki/Template:Cite_journal)</ref>[[15]](#cite_note-15)[[16]](#cite_note-16)[[17]](#cite_note-17) A study published in The New England Journal of Medicine has documented a case in which they found evidence of the Zika virus in the brain of a fetus that displayed the morphology of microcephaly.[[18]](#cite_note-18)

### Microencephaly[[edit](/index.php?title=(none)&action=edit&section=5)]

"Microcephaly" means "smallheadedness" ([New Latin](/wiki/New_Latin) *microcephalia*, from [Ancient Greek](/wiki/Ancient_Greek) μικρός *mikrós* "small" and κεφαλή *kephalé* "head"[[19]](#cite_note-19)). "Microencephaly" means "small brain". Because the size of the brain is mostly determined by the size of the head, microencephaly is implied when discussing microcephaly.[[20]](#cite_note-20)

### Other[[edit](/index.php?title=(none)&action=edit&section=6)]

After the dropping of [atomic bombs](/wiki/Atomic_bombs) "Little Boy" on [Hiroshima](/wiki/Hiroshima) and "Fat Man" on [Nagasaki](/wiki/Nagasaki), several women close to [ground zero](/wiki/Ground_zero) who had been pregnant at the time gave birth to children with microcephaly.[[21]](#cite_note-21) Microcephaly prevalence was seven of a group of 11 pregnant women at 11–17 weeks of [gestation](/wiki/Gestation) who survived the blast at less than [Template:Convert](/wiki/Template:Convert) from ground zero. Due to their proximity to the bomb, the pregnant women's [*in utero*](/wiki/In_utero) children received a [biologically significant radiation dose](/wiki/Relative_biological_effectiveness) that was relatively high due to the massive [neutron](/wiki/Neutron) output of the lower explosive-yielding [Little Boy](/wiki/Little_Boy).[[22]](#cite_note-22) Microcephaly is the only proven malformation, or congenital abnormality, found in the children of Hiroshima and Nagasaki.[[22]](#cite_note-22)

## Treatment[[edit](/index.php?title=(none)&action=edit&section=7)]

[thumb|Baby with microcephaly during a](/wiki/File:Bebê_com_microcefalia_01.jpg) [physical therapy](/wiki/Physical_therapy) session There is no cure for microcephaly.<ref name=NIH2015/> Treatment is [symptomatic](/wiki/Symptom) and supportive.<ref name=NIH2015/>

## History[[edit](/index.php?title=(none)&action=edit&section=8)]

People with microcephaly were sometimes sold to [freak shows](/wiki/Freak_shows) in North America and Europe in the 19th and early 20th centuries, where they were known by the name "pinheads". Many of them were presented as different species (e.g., "monkey man") and described as being the [missing link](/wiki/Transitional_fossil).[[23]](#cite_note-23) Famous examples are [Zip the Pinhead](/wiki/Zip_the_Pinhead) (although he may not have had microcephaly)[[24]](#cite_note-24) and [Schlitzie](/wiki/Schlitzie) the Pinhead,[[24]](#cite_note-24) who also starred in the 1932 movie [*Freaks*](/wiki/Freaks). Both these individuals were cited as influences on the development of the long-running [comic strip](/wiki/Comic_strip) character [Zippy the Pinhead](/wiki/Zippy_the_Pinhead), created by [Bill Griffith](/wiki/Bill_Griffith).[[25]](#cite_note-25)

## Notable cases[[edit](/index.php?title=(none)&action=edit&section=9)]

[Template:Unreferenced section](/wiki/Template:Unreferenced_section)

* *Triboulet* was a [jester](/wiki/Jester) of duke [René of Anjou](/wiki/René_of_Anjou) (not to be confused with the slightly later [Triboulet](/wiki/Triboulet) at the French court).
* Jenny Lee Snow and Elvira Snow, commonly referred to as Pip and Flip, were sisters with microcephaly who acted in the 1932 film [*Freaks*](/wiki/Freaks).
* Schlitze "[Schlitzie](/wiki/Schlitzie)" Surtees, possibly born Simon Metz, was a sideshow performer and actor.
* Lester ["Beetlejuice"](/wiki/Beetlejuice_(entertainer)) Napoleon Green, known on the Howard Stern Show for being "The Greatest [Wack Packer](/wiki/Wack_Pack) of All Time"

## See also[[edit](/index.php?title=(none)&action=edit&section=10)]

[Template:Portal](/wiki/Template:Portal)

* [Anencephaly](/wiki/Anencephaly) (Usually rapidly fatal)
* [Hydrocephaly](/wiki/Hydrocephalus)
* [Macrocephaly](/wiki/Macrocephaly)
* [Seckel syndrome](/wiki/Seckel_syndrome)

## References[[edit](/index.php?title=(none)&action=edit&section=11)]

[Template:Reflist](/wiki/Template:Reflist)

## External links[[edit](/index.php?title=(none)&action=edit&section=12)]

[Template:Wikimedia](/wiki/Template:Wikimedia)

* [Template:NINDS](/wiki/Template:NINDS)
* [Microcephaly Support Group](http://www.microcephaly.co.uk/)
* [The Rat People of Pakistan](http://www.telegraph.co.uk/technology/3346580/What-makes-us-human.html)
* [NINDS Overview](http://www.ninds.nih.gov/disorders/cephalic_disorders/detail_cephalic_disorders.htm)
* [Schlitzie The Pinhead](http://web.archive.org/web/20100616161524/http://www.sideshowworld.com/tgodschlitzie.html)
* [Head circumference percentile calculator](http://www.simulconsult.com/resources/measurement.html?type=head)

[Template:Congenital malformations and deformations of nervous system](/wiki/Template:Congenital_malformations_and_deformations_of_nervous_system) [Template:Authority control](/wiki/Template:Authority_control)

[Category:Congenital disorders of nervous system](/wiki/Category:Congenital_disorders_of_nervous_system) [Category:Disorders causing seizures](/wiki/Category:Disorders_causing_seizures)