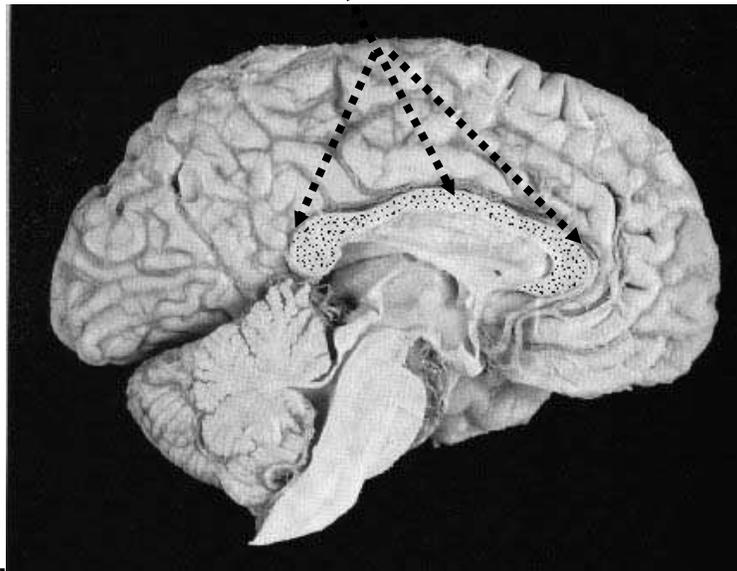


Agensis of the corpus callosum – Information for parents and carers

What is the corpus callosum?

The corpus callosum is a connection between the right and left sides of the brain, which contains many brain cells (neurones). It is shaped like an arch and is approximately 10 centimetres long in adults.

The corpus callosum



The photograph shows the left side of the brain, viewed from the midline. The front of the brain is to the right of the picture. The corpus callosum has been highlighted with dots.

How does it develop?

The corpus callosum is formed between the 7th and 17th weeks of the pregnancy, but it continues to grow and develop until later in the pregnancy, and throughout childhood.

During its formation, brain cells 'migrate' across the area between the two cerebral hemispheres and form connections with brain cells on the other side. The development of the corpus callosum takes place at a time when many other brain structures are also developing.

What does the corpus callosum do?

The brain cells in the corpus callosum send signals between the two halves of the brain (cerebral hemispheres). The corpus callosum is the largest such connection between the cerebral hemispheres, but there are also other connections. Learning, co-ordinated movements and memory all require connections between the cerebral hemispheres.

What causes abnormal development of the corpus callosum?

There are several possible causes. These are most likely to come about during the 7th to 17th weeks of the pregnancy when the corpus callosum is being formed.

- 'Migrating' brain cells may lack the chemical messengers which guide them in the right direction. This can occur because of a faulty gene.
- 'Migrating' nerve cells may be prevented from reaching their destination. This can occur because of lack of oxygen, lack of nutrients, toxic chemicals, infections or a metabolic disturbance.

How often does agenesis of the corpus callosum happen

Nobody is certain.

An estimate is around 0.5 per 10,000 births.

What do the terms 'agenesis' and 'dysgenesis' mean

Agenesis = failure to grow (implying that an organ is absent)

Dysgenesis = abnormal growth (too small; misshapen; or with a part missing)

What other abnormalities of development can occur along with callosal agenesis?

In some individuals with agenesis of the corpus callosum, nothing else abnormal will happen. In others, the following can occur:

- Other brain malformations (including brain cells elsewhere which have not 'migrated' to the right place).
- Faulty development of the intestines and kidneys.
- Faulty development of the eyes, nerves and muscles.

Agenesis of the corpus callosum may be part of a syndrome (a number of abnormalities which occur in many different individuals in a similar fashion).

In some cases it may be the result of a chromosome abnormality.

What are the consequences of agenesis of the corpus callosum?

The consequences vary hugely between different individuals

The consequences in any one individual are very dependant on which *additional* brain abnormalities are present. The consequences are not simply the result of the corpus callosum being absent or abnormally formed.

The consequences include:

- Epilepsy
- Learning difficulties (general or specific)
- Impaired co-ordination and delayed motor development
- Behavioural difficulties
- Abnormal temperature regulation and growth

In one survey of 56 cases in Britain :

- Nearly two thirds had epilepsy
- Half had intellectual impairment
- A third had a psychiatric disorder
- Nine cases were apparently normal neurologically

What can be done about it?

It is not possible to repair or replace the corpus callosum.

What can be done is to identify the medical, physical and educational consequences and to provide the correct treatment, therapy and education for these:

- Treatment of epilepsy
 - Anti-epileptic drugs
 - Steroids
 - Ketogenic diet
- Recognition and management of learning difficulties
- Management of co-ordination and motor developmental disorders

Is there pre-natal diagnosis?

This can be done using ultrasound.

The corpus callosum cannot be visualised before mid-gestation, but diagnosis of agenesis of the corpus callosum is very accurate after 20 weeks.

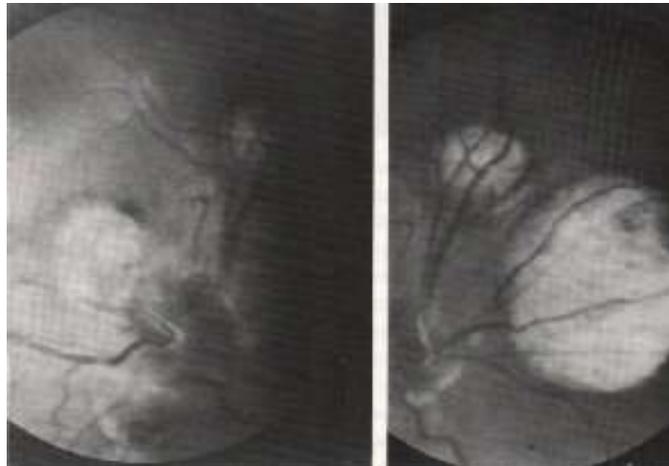
Aicardi syndrome

This condition affects girls (almost exclusively).

There is agenesis of the corpus callosum (in most cases). The retina of the eyes is abnormal. There are other brain abnormalities (cysts, collections of cells which have not migrated properly).

Many girls have infantile spasms (a form of epilepsy often starting at around 6 months of age). Most have severe learning difficulties.

In a Canadian survey of girls with Aicardi syndrome: 3 of the 14 girls could walk or crawl and 4 girls had some language ability.



Two photographs of the retina of a girl with Aicardi syndrome. The pale circles are called 'lacunae' and are found in most girls with this condition