

What are the effects of Hydrocephalus?



Hydrocephalus involves accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain, with an increase in the pressure inside the head.

Pressure

There are two sources of this pressure. One is that of the CSF itself, but a much higher pressure is produced by the heart in order to pump blood to the brain. If the CSF pressure rises, it eventually interferes with the blood supply to the brain, depriving it of oxygen and glucose which it needs in constant amounts to continue to function. Initially this causes tiredness, irritability and drowsiness, but if it progresses then loss of consciousness will result as the brain begins to shut down.

The immediate effects of this interference with the blood supply disappear if the CSF pressure is returned to normal, such as by ventricular tap or insertion of a shunt. However, in most cases the process has been continuing for some time before diagnosis of Hydrocephalus is made. During this time the interference with the blood supply leads first to a 'dying back' of the very fine blood vessels in the brain. Even this process is largely reversible if prompt action is taken, but at this time there is often insufficient clinical evidence to suspect Hydrocephalus. The next stages involve progressive damage to the actual nerve cells in the brain and to their eventual destruction, and this cannot be reversed.

One effect of raised CSF pressure may be seen in the eyes, and this is why your doctor sometimes looks for 'papilloedema'. This is caused by pressure on the blood supply to the back of the eye. It is important to realise that it may not always be present, even when the pressure is high. If CSF pressure remains high for too long, damage to the optic nerves can become permanent resulting in blindness, though fortunately nowadays this is uncommon. Another appearance, particularly in babies, is the so-called 'sunset' eye sign, where the eyes are fixed in a downward position. This is due to CSF pressure affecting important nerves running from the brain which control eye movement.

If untreated the rise in CSF pressure can cause other serious problems in the brain, unrelated to blood supply. Many of our vital functions, such as heartbeat, breathing etc, are controlled from the brain stem, a structure joining the spinal cord to the brain. Very high CSF pressure can compress this sufficiently to cause the heart and breathing to stop. Once again, this is uncommon as signs of raised pressure are usually recognised before this. A similar problem might sometimes arise, particularly in those with Spina Bifida, due to compression of the cerebellum, a part of the brain lying at the back of the head. This can also give rise to breathing, speaking and swallowing difficulties.

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Areas and Associated Functions Affected

Because of the areas of the brain most affected, functions associated with thought and learning, as well as with co-ordinated skilled movement, begin to deteriorate. The precise effects differ between individuals and are further complicated by other abnormalities, as well as by the pre-existing degrees of ability and personality of each person affected. It is not surprising therefore that while, for instance, learning disorders are common amongst those with Hydrocephalus, and their exact effects vary considerably.

Learning

There can be learning difficulties associated with Hydrocephalus such as problems with learning difficulties, behaviour, motivation and visual problems.

Much is said and written about intelligence, and particularly about IQ (intelligence quotient) in people with Hydrocephalus. In fact this is far more complicated, and a good deal less informative, than many believe. The IQ is made up of several components which can be thought of as verbal and non-verbal, or performance-related tests. People with Hydrocephalus generally score better on verbal IQ than on performance IQ and this is thought to reflect the distribution of nerve damage in the brain as described above. Certainly, during periods of rising CSF pressure, such as in untreated cases or when a shunt is blocked, the effect on performance IQ is more marked. Generally speaking, people who have had Hydrocephalus since birth or childhood have, as a group, a lower average IQ than a comparable group without Hydrocephalus, but it is important to realise that there is a wide range in each group, and some people with Hydrocephalus have very high scores.

Practical Implications

Hydrocephalus can also result in subtle effects, giving problems with co-ordination, motivation, organisational skills and language. Physical effects such as visual problems, or early puberty in children, may also occur.

The practical implications of these features of Hydrocephalus are that there may be subtle problems of co-ordination of hand movements with what the person sees, as well as a degree of clumsiness, which make it difficult to perform certain tasks or do certain jobs. With regard to learning in the home or to education in school, there may be real problems with concentration and reasoning which require a sympathetic but skilled approach. For instance, it will often be necessary to teach simple everyday tasks like getting out of bed, washing one's face, dressing and going downstairs as separate short items rather than all at

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once, and to keep them consistent and repetitive. This does not indicate 'stupidity', but is caused by damage to the nerves in the brain which normally allow us to learn very quickly how to do a complex series of things. Much can be done to help, and the following chapters of this book give parents and teachers practical advice on how to help children overcome many of these difficulties. Professional advice should be sought where needed.

Many of these effects can be reduced through teaching strategies or with treatment where relevant.

Psychological Development

Psychological development in children and adolescents with Hydrocephalus may proceed normally, but sometimes the changes associated with puberty (breast development, body hair growth etc) appear much earlier than expected, and the intrusion of psychological aspects of sexual development into a mind which is emotionally still very immature can cause distressing problems (see 'Precocious puberty', Again, specialist advice should be sought if necessary. Other effects of Hydrocephalus may also be seen, and some of these are difficult to explain. For instance, some people are very seriously distressed by every-day noises such as vacuum cleaners or washing machines.

Sensitivity to Noise

Many people with Hydrocephalus are very sensitive to sudden high-pitched sounds or very loud noises, e.g. amplification. Young children react by crying and may become very distressed although many become less sensitive over time. Some adults report feeling sensations in the shunt and others have an echoing feeling in the head, others feel panic, nausea and may burst into tears.

Seizures

Approximately one third of people with Hydrocephalus have seizures at some time in their lives. A rise in intra cranial pressure due to shunt blockage may trigger an Epileptic fit. Fits sometimes occur after shunt revision. It is often just an isolated incident, but some people go on to develop Epilepsy. Epilepsy is usually treated with anti-convulsing drugs and is the same for people with or without Hydrocephalus.

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Premature Puberty

Some children with Hydrocephalus may develop early puberty. It is seen more often in girls than in boys. Preparation of the child for the onset of periods and sexual development needs to be handled sensitively.

Eye Problems

Eye problems may be the first sign of raised pressure in the brain or shunt blockage, so it is important to monitor the eyes. Visual assessments and ocular assessments, which monitor the eye movements and examine the back of the eye, are recommended.

There is a high incidence of eye problems in patients with Hydrocephalus, such as strabismus (squint), nystagmus (fine wobble of the eyes), papilloedema, (swelling of the optic disk), optic atrophy and blindness. All Hydrocephalic children are at risk of losing vision and developing a squint. Squints can cause problems with judging distances, speed of approaching vehicles etc.

Language

In some children with Hydrocephalus their ability to use language is often ahead of their ability to understand it. Their vocabulary can be good because they are able to imitate what they hear. If they do not understand fully what is said their response may be inappropriate. A child with a language problem will pick out words they understand and guess the rest or give a stock answer.

Speech

If the child has a problem with forming sounds he/she may need to be referred to a Speech Therapist.

Weak Upper Limb Control and Hand Function

Problems with upper limb control and hand skills are common in people with Spina Bifida and Hydrocephalus. The majority have weak muscle power in their upper limbs. It is important to encourage the use of both hands, especially if one hand is weaker than the other. Loss of sensation in the hands results in an inability to discriminate between differences in water temperature or the temperature of objects such as radiators, kettles

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etc. Problems with fine finger movements are shown in everyday tasks such as fastening buttons, threading needles, catching balls, screwing lids on jars and using scissors, as well as handwriting. For many everyday tasks and handwriting, it is necessary to stabilise trunks and shoulders and maintain a good sitting position. The non-dominant hand should be used as a support e.g. to hold the copybook as the child uses the dominant hand to write.

Visual Perception

People with Hydrocephalus often have problems with visual perception. Although they recognise objects, they find it difficult to understand their position and relationships. A squint or other eye problems can exacerbate the problem. Some experience loss of depth perception or have difficulty in judging distance or speed. There may be a difficulty with scanning visual images, with consequences for reading, writing and drawing.

Difficulty in discriminating between different shapes has implications for learning to read and write, also problems with shape, size, direction, volume and position can cause difficulties with maths and practical tasks. Figure ground discrimination i.e. identifying an object from its background, may show up in problems with crossing a road, maps, diagrams and artwork in school.

Visual perception difficulties also means that judging slopes, height of kerb, width of doors or space in a room may prove problematic. It also affects placing objects accurately i.e. a glass left near the edge of a table and even feeding - difficulty in getting spoon or cup accurately to the mouth. Dressing and undressing e.g. getting clothes on inside out, upside down etc may be difficult too. Sometimes people with Hydrocephalus are not aware of signals given by facial expressions in others.

Perceptual difficulties are not the only problems associated with Hydrocephalus. Some people with Hydrocephalus may have problems with decision-making, logical thinking, organisational problems, and inability to follow verbal instructions, short-term memory difficulties and passive behaviour. All of these have major implications for adult life.

Spatial Awareness

Spatial awareness is the ability to understand the surrounding space and judge distance, height, width, size, volume. Problems with spatial and visual perception are inter-related. It can affect the way people move about, e.g. a wheelchair user may bump into tables, graze doorways or clip people's heels. Fear of tilting a wheelchair backward to climb a kerb or fear of the drop may also be apparent. People with Hydrocephalus may have a

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fear of being left in a room alone, of venturing outside, or of long corridors. Manual tasks might be carried out poorly e.g. matching buttons to buttonholes, getting an arm into a sleeve, laying a table, making a bed. Their handwriting might also be quite poor and illegible - letters uneven or poorly spaced, mixture of upper and lower case, due to visual perception dysfunction.

What does this mean?

Reading a catalogue of the effects of Hydrocephalus can be very alarming. However, it should be realised that some people with Hydrocephalus may have very few of these problems. Also, many of those which have been described are found either in untreated Hydrocephalus or when the treatment fails, and when successful treatment has been promptly introduced they often improve or sometimes disappear. On the other hand, the more subtle learning and reasoning problems are usually present in some degree and are very important where a child's development and education are concerned.

Many of these effects can be reduced through teaching strategies or with treatment where relevant. For further information on specific areas of concern see our Continence, Learning & Teaching, Health Body/Health Mind and Living Your Best Life sections at www.sbhi.ie

It must be stressed that the effects of Hydrocephalus vary from one individual to another and some people will have very few, if any, problems.