Hydrocephalus

Hydrocephalus: The Battle for Shunt Placement

It is oftentimes blatantly obvious that children born with hydranencephaly have heads that are disproportionately large at birth, one of the first signs that there is an obvious neurological problem especially in utero. Yet doctors do not always believe that any sort of medical intervention, such as shunt placement to drain excess fluid, needs to be explored. In fact, many parents are led to believe that it is a senseless procedure considering the prognosis and the possibilities of these children having an adverse reaction to the anesthesia, amongst other risks of any type of surgery. In other cases, the diagnosis of hydranencephaly becomes overlooked or misdiagnosed altogether, replaced by a diagnosis of severe hydrocephalus with the concept of fluid pushing the cortex up around the inside of the skull rather than being non-existent (Kaneshiro, 2010). Regardless, doctors view the possibility of not surviving the surgery as a greater risk than the chances of it making a difference to the quality of life they live. More often than not, this is simply not a true sentiment to grasp as acceptance for a parent to a child who appears to be in discomfort.

Not all children with a diagnosis of hydranencephaly experience hydrocephalus, a build-up of cerebrospinal fluid or “water on the brain” (Kaneshiro, 2010), at birth. Some present this symptom later in their life, and others never at all. Still other children actually display the opposite with a condition known as microcephaly, where the cranial circumference actually presents as much smaller than average due to brain malformation (NINDS, 2008). In any instance, focus should be on ensuring that the child lives a comfortable life. Regardless of diagnosis and associated prognosis, your child has rights to appropriate medical treatment with your full involvement as their parent. Since many doctors have never treated a child with hydranencephaly, it is important that you possess extensive knowledge of your child and share that knowledge with those who will be caring for them.

In cases of hydranencephaly, children are missing a majority of their essential brain components. The flow of cerebrospinal fluid (CSF), the fluid that transports nutrients to and waste from the brain is no longer regulated. In ideal circumstances, CSF generally does its designated job, afterwards finding itself reabsorbed in to the bloodstream, but build-up exists when either too much is created or a full or partial blockage exists (Kaneshiro, 2010).
At birth, hydrocephalus is easily identifiable by an abnormally large head circumference. Other signs are a bulging fontanel (soft spot), downward gazing (sun setting) eyes, varying degrees of seizure activity, obvious spaces between sutures (skull bones), lethargy, vomiting, abnormal reflexes, and irritability. For children that later develop hydrocephalus with hydranencephaly, the symptoms are similar. A confirmed diagnosis is generally reached via CT scan, MRI, or other imaging devices (Kaneshiro, 2010).

The goal of treating hydrocephalus is not only for comfort, but also to prevent further damage to the existing brain components in children with hydranencephaly. Treatment is achievable through placement of a shunt, flexible tubing that allows flow of the excess CSF to another part of the body for re-absorption. In some cases where overproduction of CSF is detected, the part of the brain that creates CSF can be cauterized or removed all together, though not ideal in most cases (Kaneshiro, 2010).

Since shunt placement proves to be the ideal treatment for hydrocephalus, this topic is further detailed. There are different types of shunts, essentially they all are compiled of three main components: the shunt catheter (small, flexible tubing), connected to the reservoir or valve (which controls the flow of CSF), and the lower shunt catheter that drains in to the re-absorption site (which is most generally the abdomen). A VP shunt (ventriculo-peritoneal shunt) is that which drains in to the abdomen, while ventriculo-pleural drains in to the lung, the ventriculo-atrial (VA) drains in to the heart, and other not-so-common shunts can be directed to the gallbladder or bladder. The flow of drainage can be either fixed at low, medium, or high or programmable by your neurosurgeon with a magnet as necessary (NHF, 2007).

Though there is a whole host of pros to shunt placement surgery for a child with hydranencephaly, the biggest con is that it is a lifetime commitment. Shunts do not last forever; they can malfunction or fail, thus requiring a revision. Infection, blockage, calcifications or catheter breakage, broken or stuck valves, and separation from shunt components can all be reason for malfunction. Other reasons, such as lengthening of the catheter and/or flow-rate change, can be easily avoided with extra tubing coiled within the re-absorption site to compensate for growth of the child and placement of a programmable shunt for easy adjustment to flow-rates (NHF, 2007).

Many parents are “fortunate” that the build-up of CSF is rather fast, thus noticeable, while many times it is nearly undetectable. One thing to remember is that hydrocephalus is capable of causing great amounts of pain and discomfort, placing it as one of the main causes of irritability in infants with any kind of brain trauma. Surgery itself carries risks, though most
endure surgery for shunt placement without any complications. A tiny hole is drilled into the skull and the catheter is run from the head behind the ear, down the neck and chest, and into the abdominal cavity (Allanach, 2010)... only 2 teeny-tiny scars are apparent, one on the head and one on the belly, and you will barely feel the tube at all! The actual shunt site is a bit prominent at first, but it becomes much less obvious after all of the swelling goes down.

The visual aftermath of surgery is a warning I wish had come along in the guidelines from the surgeon. In cases of extreme CSF build-up, upon shunt placement the head becomes comparable to a bowl of gelatin, complete with the jiggle upon slightest repositioning. The cranial bones, sutures, protrude while the fontanel has become cavernous with the extreme decrease in pressure. Eventually, with persistence and constant repositioning and awareness, the little head will become much more cosmetically appealing. From a personal standpoint, shunt surgery saves these little lives and allows them to thrive and live a much greater quality of life despite the associated grim prognosis.

References


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