The following article was written for the Little People of British Columbia Association by Dr. Paul Steinbok HB FRCS (C), an associate of Dr. Judith G. Hall's at Children's Hospital in Vancouver, B.C. Dr. Steinbok has treated many members of LPBC for hydrocephalus and has conducted studies accordingly. He comes highly recommended by Dr. Hall.

Sincerely, Muriel Reid, President, LPBC.

Hydrocephalus in Achondroplasia

Children with achondroplasia are known to have enlarged heads which in most instances is related to the fact that the fluid cavities in the brain (ventricles) are enlarged. One of the major causes of ventricular enlargement is hydrocephalus, a situation in which the pressure within the ventricles is higher than normal. Hydrocephalus often manifests itself early in life with increasing head size, and later in life with problems such as headaches, vomiting, and obvious evidence of brain disfunction. In children with achondroplasia there has been much controversy about the significance of the ventricular enlargement because most children do not exhibit headaches, vomiting, lethargy, and other symptoms which one associates with hydrocephalus. Furthermore, it has been felt that in children with achondroplasia, the ventricular enlargement stabilizes with time and does not lead to significant problems.

We have been concerned about this approach to children with achondroplasia because it is known from other situations in which tydrocephalus exists, that it is possible to have more chronic hydrocephalus which can result in abnormal function of the brain, evidenced only by a decrease in the level of intelligence and functioning at school, or by mild intermittent headaches. It was our feeling at the British Columbia Children's Hospital that if hydrocephalus acutally did exist in achondroplasia, and if the pressure within the ventricles was higher than it should be, then there might be persistent chronic damage occuring to the brain, and that this would not be an ideal situation for anyone. Because of this concern, we studied a number of achondroplastic children with big ventricles. Firstly, we measured the pressure within the ventricles by putting a small tube into the brain and hooking that up to a monitoring device. We were able to show in five children in whom this was done, that the pressure was elevated. While the tube was still in place, we took the opportunity to inject a dye-like material (contrast), and also some radioactive material into the ventricles to see if there was any blockage to the flow of spinal fluid within the brain that might account for the big ventricles and hydrocephalus. With this test, we were able to show that there was no blockage. This indicated to us that the problem causing the hydrocephalus in achondroplastic children might be related to a decrease in the absorption of spinal fluid. Spinal fluid is normally absorbed over the top of the brain into the big veins that drain the brain. It was postulated that perhaps the pressure in these big veins was too high, and that this was inhibiting the absorption of spinal fluid. It was thought that this high pressure could be produced by a blockage to the flow of venous blood at the base of the skull, which is known to be very small in children with achondroplasia. By putting small catheters into the venous system via the groin, and injecting dye into jugular venis which drain the brain, we have shown that there is narrowing of the veins at the base of the skull where the skull meets the neck. We have also been able to show that this does not cause an increase in the pressure in the veins draining the brain. In two children, it appeared also that there might be a constriction or narrowing of the jugular vein as it went from the neck into the chest, which is also small in children with achondroplasia.

Based on the above studies, we have concluded that in children with achondroplasia, hydrocephalus is quite common, and is likely related to obstruction to the flow of venous blood from the brain. We believe that this is a significant problem, and have attempted to treat this by shunting fluid from the ventricles to the peritoneal cavity in the abdomen. In this way we have attempted to reduce the pressure within the brain to normal, and thus prevent further damage from occurring to the brain. Our initial results have been encouraging.

We believe that it is important that all children with achondroplasia be followed by measurements of the head circumference. Those children whose heads are particularly large are more likely to have hydrocephalus, and are more likely to benefit from any type of shunt procedure. More importantly perhaps, is the approach to newborns with achondroplasia, because these children may be born with normal ventricles and may develop hydrocephalus only later on in the first year or two of life. It is easy to follow these newborns with repeated ultrasounds of the brain to look at the size of the ventricles, and if the ventricles start to increase in size, it may be useful to consider treatment earlier rather than later.

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NO it isn't Christmas in June but it is the first time space was available for this picture of the Flinida Mini Gators Party