**Introduction**
"Jon" is a 4-year-old boy with Wolf-Hirschhorn Syndrome. He is on medications for a seizure disorder but still has occasional seizures (especially when he's ill), along with myclonic jerks. He was born with cleft lip and palate which have been repaired, but still has a poor suck and swallow and receives continuous tube feedings through a GJ tube (a tube that enters the stomach and delivers food directly to the small intestine) for 20 hours/day.

Jon is an extremely social child with a very interactive personality. He has good receptive language and is now communicating through a variety of signs and gestures. He enjoys music and singing and will do gestures to several different songs He also makes lots of vocalizations, but only has a few recognizable words.

He receives weekly special education, speech therapy, OT, and PT, and has recently begun feeding therapy

Jon is beginning to make choices and show preference through signs. He can manipulate a variety of objects with both hands and place objects into containers. He can transition into sitting and is beginning to independently move around his environment by rolling. He can stand at a support surface for several minutes with close supervision and his PT and family are exploring walkers and gait trainers.

While he has delays in all areas of development, Jon has made a lot of great progress. This is especially impressive when you consider that his parents were told by doctors that he wouldn't live to see his first birthday.

**What is Wolf-Hirschhorn Syndrome?**
Wolf-Hirschhorn Syndrome is a very rare genetic condition that occurs in 1 per 50,000 births. Children with this condition can have some or all of conditions listed below. This condition is caused by a partial loss of material from the short arm ("p") of chromosome 4. *The amount of chromosome material that is lost determines the severity of the child's condition.* A diagnosis is made by blood tests. Most (87%) children with this condition have it because of a new chromosomal defect. Unless one of their parents is a carrier for the disorder, the risk of having another child with Wolf-Hirschhorn syndrome is very low.

**What Do Children with this condition look like?**
Common Physical traits of children with Wolf-Hirschhorn Syndrome include:

- Small body size when they are born and slow growth throughout life
- Small head size
- Wide set eyes
- Broad or beaked nose ("Greek Warrior Helmet" appearance)
- Cleft lip and/or palate (roof of the mouth)
- Low set and unusually shaped ears
- Low muscle tone and muscle weakness

Other physical findings can include:

- Eye defects
- Bone abnormalities (Scoliosis, abnormal numbers of ribs, hand deformities)
- tooth abnormalities
- small chins

A Boy And Girl With 4p-* A Greek Warrior Helmet*

**What Common Medical Concerns Occur with this Condition?**

Children with this condition often have a number of the following medical concerns:

- Seizures
- Hydrocephalus (excess fluid in the brain)
- Abnormal brain development (agenesis of corpus callosum)
- Heart defects
- Feeding difficulties
- Hearing impairment
- Abnormally shaped kidneys

**What Behavioral/Developmental Concerns Occur with this Condition?**

Children with Wolf-Hirschhorn syndrome commonly demonstrate:

- Moderate to severe development delay
- Moderate to severe mental retardation
- Hand ringing/hand flapping behaviors
- Rocking, head-shaking
- Moderate to severe speech impairment
- Note: speech is usually absent and comprehension is limited to simple orders or to a specific context, but this may be related to the size of the gene deletion as there have been reports of some children with small deletions who have been able to communicate through speech.

**What is the Life Expectancy of Children who have this condition?**
In the past, it was generally thought that these children had shortened life spans. However, it is now evident that individuals with WHS are capable of living into their teens and beyond, especially those who have milder forms of the condition because of smaller chromosome defects. Some studies have reported that 1 out of 5 children will die within the first 2 years of life. For others it is between 18 and 34 years of age. Again, *there is a statistically significant relationship between size of the gene defect on chromosome 4 and the child's overall risk of death.*

**Are There Any Available Treatments for this Condition?**
Since no treatment exists for the underlying genetic disorder, treatment for Wolf-Hirschhorn syndrome focuses on the symptoms present. For example, a seizure disorder would be treated with medication, and difficulty eating or swallowing might require a gastrostomy feeding tube. Medical evaluations should include an eye exam, hearing screening, heart exam, ultrasound of the kidneys, EEG and swallow study. Physical, occupational and speech therapy can be helpful in addressing the child's mobility, self-care, play, and communication needs. Coordinated multidisciplinary care is the best approach.

**Where Can I Get More Information?**
4p- Support Group - http://www.4p-supportgroup.org/

Online Threaded Discussion List about 4p- http://www.4p-supportgroup.org/forum/

Wolf-Hirschhorn Support Group UK - http://www.whs.webk.co.uk/

Aaron Willock's page (from the parents of a boy with Wolf-Hirschhorn Syndrome)
http://www.willock.net/arran/

*Children's images courtesy of *The GAPS Index* http://www.icomm.ca/geneinfo/wolf.htm
*Greek Helmet image courtesy of *Great Commission Illustrated Books*
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