

**SECOND ANNUAL NIH CONFERENCE ON HOLOPROSENCEPHALY
FAMILY SESSIONS: TUESDAY, APRIL 9, 2002
SESSION III: MANAGEMENT**

Motor Issues and Current Treatment

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Patients with holoprosencephaly (HPE) are commonly affected by some type of motor disorder. Actually, all patients with HPE that have been evaluated at the Carter Centers have some degree of disability due to a motor impairment. This motor disorder can be described as a form of cerebral palsy (CP). Cerebral palsy has been defined as “A collection of non-progressive disorders that manifest as abnormalities of motion and posture and result from a central nervous system injury sustained in the early period of brain development, usually defined as the first 3 to 5 years of life”. Therefore, HPE is one of many causes of cerebral palsy. The motor disorder in patients with CP and in HPE may be very complex resulting in weakness, poor motor planning, coordination problems, abnormal tone, involuntary movements and sensory deficits. Although the definition of cerebral palsy implies that the disorder is not progressive, the manifestations of this non-progressive brain disorder may actually change due to growth and maturation of the child. The effect of muscle imbalance around joints due to weakness and abnormal tone may eventually produce joint dislocations, soft tissue and muscle contractures and bone deformities adding more disability to the patient.

Out of 35 children with HPE over 18 months of age, 37 % sit independently, 9% walk with some assistance and only 11% walk independently. Out of 46 patients older than 1 year of age, 72% can hold objects in their hands, 41% can reach for objects and only 11% have normal hand function. The degree of motor impairment is directly related to the severity of the brain malformation. Thus, patients with alobar HPE have the most severe motor deficits while patients with lobar and MIHF HPE are the least affected.

References:

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