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**Functional outcomes of Infants with Chiari II Malformation with Tracheostomy and Home Ventilator Dependence**

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Chiari II malformation has the incidence of 1 in 1000 live births.

Characteristic features include peaked midbrain, downward displacement of cerebellar tonsils and vermis, and spinal myelomeningocele which can be associated with hydrocephalus.

Mortality is 50-70% if not recognized early.

Survivors have a high morbidity and develop neurologic abnormalities and significant developmental delays.

A subset of infants with Chiari II malformation may develop central and obstructive apneas that may require long-term ventilator support through a tracheostomy.

Objective- To describe the short-term functional outcomes of patients with Chiari II malformation who are ventilator dependent at home through tracheostomy.

IRB approved retrospective study.

Over the 8-year period (2010-2018), 85 infants were discharged from the hospital with Chiari II malformation.

Of the 85 infants, 9 (10.5%) received tracheostomy for home ventilation due to vocal cord paralysis and central apnea.

Tracheostomy was placed within 3 months (range 1.5-3 months) of life.

Of these 9 infants, 1 (11%) was successfully decannulated at 2 years of age, 3 (33%) died, and 5 (56%) remained tracheostomy and ventilator dependent.

By the age of 3 years of age, all surviving infants were non-ambulatory without assistive support and had significant speech/language delays.

8 (89%) of the 9 infants continue to require G-tube feeds for nutrition.

These findings will provide additional data regarding the short-term functional outcomes of a subset of infants with Chiari II malformation who are ventilator-dependent at home. Furthermore, awareness of these findings will help health care providers and parents to make well-informed care decisions for these medically complex children.