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

Abbigayle Carlson

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

Submitting/Presenting Author (must be a trainee): Abbigayle Carlson
Primary Email Address: abbigaylecarlson@kansascity.edu

X Medical Student
☐Resident/Psychology Intern (≤ 1 month of dedicated research time)
☐Resident/Ph.D/post graduate (> 1 month of dedicated research time)
☐Fellow

Primary Mentor (one name only): Gangaram Akangire, MD
Other authors/contributors involved in project:
Addie Begley, ARNP, Charisse Lachica, MD, Winston Manimtim, MD

IRB Number: Through CIPD (Center for Infant Pulmonary Disorders)

Describe role of Submitting/Presenting Trainee in this project (limit 150 words):

Presenting author was involved in data abstraction. She prepared the data excel sheet for data analysis. She was involved in data analysis and writing, reviewing and editing the abstract.

Background:
Chiari II malformation is relatively common with an incidence of about 1 in 1000 live birth and is associated with reduced life expectancy. Characteristic features are beaked midbrain, downward displacement of the tonsils, and cerebellar vermis, and spinal myelomeningocele that can be associated with hydrocephalus needing ventriculoperitoneal shunt (VP shunt). Mortality is about 50-70% if not recognized early. Survivors have high morbidity and develop neurologic abnormalities and significant developmental delays. Long-term outcome is poorly understood.

Objective:
Not much has been published that describes the outcome of infant with Chiari II malformation that received tracheostomy for long-term ventilator support. Our objective was to describe the functional outcomes of a series of 9 patients with Chiari II malformation with tracheostomy and ventilator dependence over 8 year period and 3 years after birth.

Methods:
A retrospective study of infants with myelomeningocele and chiari II malformation who received tracheostomy for long-term home ventilation was designed. Study period included 2009 to 2018. Infants with no tracheostomy were excluded from the study. A 3-year analysis of survival and decannulation from tracheostomy, length of ventilator dependence and

gastrostomy feeds (GT) for nutritional needs, urinary catheter dependence and ability to ambulate was assessed.

Results:

Over 8 years (2010-2018), a total of 85 infants with Chiari II were discharged from the hospital. Of these, 9 (10%) had tracheostomy for home ventilation. Seven (7) infants were born at term and 1 was premature born at 32 weeks and 1 was late preterm born at 35 weeks. Cohort included 4 males and 5 females. Myelomeningocele repaired and ventriculoperitoneal shunt (VP) shunt was placed at median age of 1 day (range 1-5 days) and 5 days (1-46 days) respectively. Tracheostomy was placed within 3 months of age (1 – 3 months) due to central apnea and vocal cord paralysis. All infants were provided chronic ventilation at parent's request. Current ages range from 3 to 9 years. Of the 9, 1 was decannulated, 3 died (Figure 1) and 5 remain tracheostomy-dependent. Of these 5, 2 weaned off the ventilator, 2 receive nocturnal ventilation, and one was ventilator-dependent 24 hours/day. By the end of 3 years after birth, all 9 infants were non-ambulatory without support, 8 infants were on GT feeds, one infant was successfully decannulated and 5 infants were still on ventilator support.

At 3 years of age, all infants were non-ambulatory without support and have speech/language delay and receiving physical, occupational and speech therapies.

Conclusion: In a series of 9 infants with Chiari II malformation with tracheostomy, only 1 was successfully decannulated. All continue to have complex medical needs with significant functional delays. These results underscore the ongoing morbidities, neurodevelopmental impairments, and risk for mortality faced by this vulnerable population. These findings will be a good addition to the literature and will be helpful in explaining the prognosis of infants with Chiari II malformation with tracheostomy to parents. This will further help parents in taking a informed decision to pursue tracheostomy.

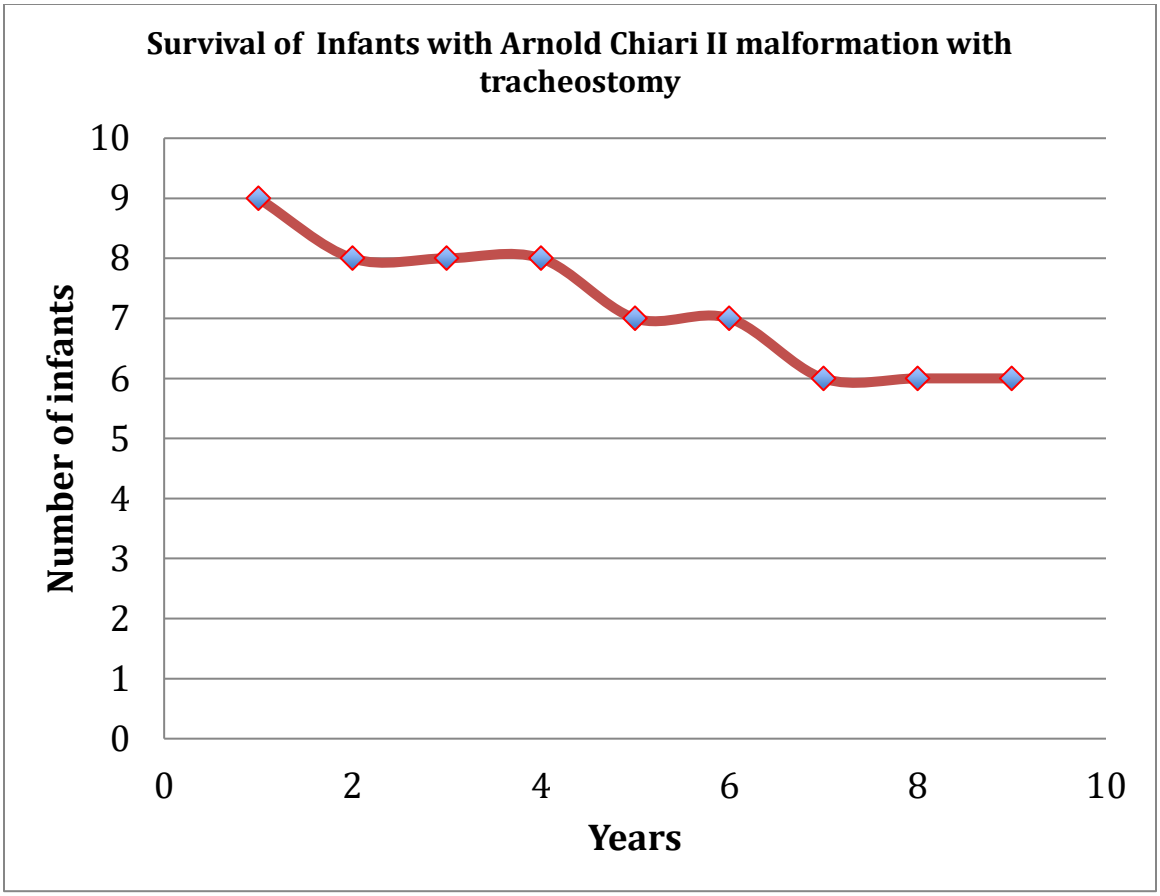

Figure 1: Survival curve for the entire cohort