Little Folk Strokes: Current Questions

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Pediatric stroke (defined as stroke occurring between 1 month and 18 years of age) is more common than many highly visible diseases that occur in adults and, as such, is an under-recognized problem. The incidence is at least 2–3/100,000 children per year with a 10% mortality rate. This represents ~100 new pediatric strokes every year in New York City alone. In pediatric stroke, any deficit results in cognitive and motor disabilities that persist over decades during the remainder of the affected person's life. Despite this, there is little public recognition and there are no clinical trials except in sickle cell disease- (SCD) associated stroke.

Newborn stroke, which occurs anytime between in utero through 1 month of age, has an even higher incidence than childhood stroke (4–5/10,000 live full-term newborns). Most newborn strokes present with seizures and probably 50% are clinically silent, suggesting that the incidence of newborn stroke is at least 1/1000 live-born full-term infants. This number does not include the higher incidence of premature birth associated hemorrhage and ischemic disease common in premature infants. Newborn stroke does not recur and an etiology is found in ~50% of cases. There is little data that conditions at delivery play a significant etiological role and coagulation defects are likely more commonly found than seen in older stroke patients. There are few studies other than case series and no treatment trials.

There is no evidence-based treatment of pediatric stroke in the acute phase and no safety and efficacy data. Publicizing that strokes occur in children will decrease time to the hospital, so that attempts at acute treatment can be planned. There are no current standards, nor plans on developing standards, for children. Additional protocols need to be developed for the treatment of children in the acute situation detailing the higher incidence of premature birth associated hemorrhage and ischemic disease common in premature infants. Newborn stroke does not recur and an etiology is found in ~50% of cases. There is little data that conditions at delivery play a significant etiological role and coagulation defects are likely more commonly found than seen in older stroke patients. There are few studies other than case series and no treatment trials.

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In this issue of CNS Spectrums, a series of articles brings to the fore some of the key current questions and issues in newborn and pediatric stroke. Akila Venkataraman, MD, and colleagues present a case series of newborn stroke. Most patients have left middle cerebral artery infarcts and some have multifocal strokes. It is likely that the location of stroke may help identify etiologies and even timing of stroke. Possibly, certain stroke types are more likely prenatal whereas others occur closer to the time of birth. While the clinical presentation of seizure in the newborn period and corresponding imaging characteristics of the stroke may suggest that a stroke occurred recently, in patients with silent infarcts diagnosed later in infancy with the development of early hand preference, the stroke may have been remote to the time of birth. These questions are only posed by the authors but should be included in studies going forward. The questions have medico-legal implications and more importantly are crucial before any treatment trials are considered.

Fenella J. Kirkham, MB BChir, and Alexandra M. Hogan, PhD, detail the known risk factors for stroke in older children. The data is obtained from many of their own studies at a large tertiary teaching hospital in London. They opine that most of the data and information available is from case series at tertiary centers. Despite this lack of clinical studies, the information that the authors bring is substantial and is a first step to a better understanding of cerebrovascular disease in childhood.

Ruth D. Nass, MD, and Doris Trauner, MD, have a longstanding interest in cognitive outcome of stroke particularly stroke in the newborn. They explain how the timing of stroke may affect language outcome and the location of stroke affects language in different ways depending on the timing. Subtle language abnormalities were, until recently, overlooked and represent an under-recognized life-long problem in those affected. Strokes in those <2 years not only affect language and motor functions but impact on behavior, attention, and psychiatric functions. The study of newborn stroke will prove insightful into our understanding of the timing of development and anatomic correlates of normal and abnormal behavior. Both the timing and location of stroke impact on behavior, language, and motor function in different ways.

Mehari Gebreyohanns, MD, and Robert J. Adams, MS, MD, describe the importance of clinical trials and discuss primary stroke prevention in SCD. Dr. Adams
was the principal investigator in the Stroke Prevention Trial in Sickle Cell Anemia, which found that abnormal elevation in Doppler velocity studies of the middle cerebral artery predict an elevated (10-fold) stroke risk and the initiation of transfusion therapy decrease the risk by >90%. This represents the most compelling example of primary stroke prevention known. The authors also discuss the importance of Doppler studies in SCD and challenges with the utilization of Doppler in this population. Silent infarction and its correlation to cognitive deterioration is highlighted.

Sharon Friefeld, PhD, and colleagues report a major reduction in quality of life following pediatric stroke in both parent-proxy and child self-report Health-Related Quality of Life scores compared with normal children or those with other chronic health conditions. Of greatest concern for parents and children is the effect of stroke on school and its impact on emotional and social functions. The authors show that the severity of neurological deficits after stroke is the most significant predictor of poor quality of life.

Here, many questions in newborn and pediatric stroke are posed and a few are answered, especially in SCD. Hopefully, over the next 10 years, sufficient information will be available from currently ongoing epidemiological and future clinical trials to answer the many questions posed here. Suffice it to say, clinical questions require clinical trials.

REFERENCES