Perinatal Arterial Ischemic Stroke

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During childhood, the neonatal period is the time when the risk of ischemic or hemorrhagic stroke is highest. The focus of this chapter is perinatal arterial ischemic stroke (PAIS), defined as "a cerebrovascular event occurring during fetal or neonatal life, before 28 days after birth, with pathological or radiological evidence of focal arterial infarction of brain." Discussion of perinatal cerebral sinusovenous thrombosis is included in Chapter 3, and discussion of intracerebral hemorrhage in neonates can be found in Chapter 4.

In recent population-based studies, the incidence of PAIS has been reported as 13 to 23/100,000 births/year in term and near-term infants. Rates as high as 1/2300 births have been reported in smaller studies. The incidence of PAIS is comparable to that of large vessel ischemic stroke in adults. Not only is PAIS relatively common, but the clinical impact is significant. PAIS accounts for 30% of cases of congenital hemiparesis in term infants, making it the most common cause of hemiplegic cerebral palsy in this population.7 In this chapter, the typical clinical presentation and risk factors of PAIS will be reviewed. Recommended evaluation will be discussed, and representative neuroimaging will be presented. Treatment and outcome will also be reviewed.

Clinical Presentation

The majority of neonates with PAIS are diagnosed within the first 28 days of life with an acute stroke. In one population-based study that included children with an acute presentation as well as those with a delayed presentation, 58% came to attention during the neonatal period. In an international registry, the vast majority (87%) were diagnosed within the first week of life. Among term infants, clinical seizures are the most common presenting symptom, appearing in approximately 70%, and 12% of neonatal seizures are due to PAIS. Nonspecific symptoms such as abnormal tone, altered level of consciousness, and respiratory or feeding difficulties are also common. Focal neurological signs occur in the minority. Stroke affects the left hemisphere more often than the right, and most perinatal strokes involve the middle cerebral artery (MCA) territory.

Acute PAIS also occurs in preterm infants, though less is known about this subgroup. In one series, the majority (83%) came to clinical attention because of apnea or other respiratory difficulties, and only 30% presented with seizures. In a population-based study that included term and preterm infants, acute stroke diagnosis was prompted by an incidental abnormality on routine surveillance head ultrasounds in all but one preterm infant. PAIS was then confirmed by additional neuroimaging. As in term infants, preterm infants were more likely to have PAIS on the left side, and MCA location was most common.

More than one-third of infants with PAIS do not have recognizable symptoms in the neonatal period, and diagnosis is made months to years later. Most often these children come to medical attention because of early hand preference indicating an emerging hemiparesis. In other cases, seizures or developmental delays prompt neuroimaging, which reveals a chronic arterial ischemic stroke.