Neurophysiological studies in infants with Vein of Galen aneurismatic malformation (VGAM): Bambino Gesù Children Hospital experience in the last two years

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Objective. Vein of Galen aneurysmal malformation (VGAM) is a rare cerebral vascular congenital anomaly due to intracerebral arteriovenous shunt across the remnant of an embryological structure, known as Markowski’s vein, affecting less than one birth in 25,000. VGAM represents the most frequent cerebral vascular anomaly in the pediatric population. Our goal was to evaluate neurophysiological features such as sensitive, visive and auditive responses in newborns with VGAM.

Materials and Methods. In the past two years, from April 2020, at Bambino Gesù Pediatric Hospital in Rome, we have taken in 11 neonates with VGAM, 9 of them with prenatal diagnosis. We have subjected them to non-invasive neurophysiological exams including sensory evoked potentials (SEP). These are recorded from the central nervous system following stimulation of sense organs and include somatosensory (SSEP), visual (VEP) and brainstem auditory evoked potentials (BAEPs).

Results. In our cohort SSEP gave pathological findings in 6 neonates, specifically we found asymmetric responses in 5 patients and a total lack of response in one of them. Visual evoked potentials were altered in 3/8 while BAEPs resulted asymmetric in only one in 8 patients.

Conclusions. VGAM remains one of the most challenging vascular anomalies in pediatric patients, clinically and therapeutically. They have a higher risk of poor long-term outcome in terms of neurological development and neurophysiological impairment. In our opinion a complete neurophysiological functions assessment is essential to begin a personalized rehabilitation treatment.