Rare Vein of Galen malformations (VOGM) or median prosencephalic arteriovenous fistulas occur about one in 3 million but related to the high mortality rate in spite of the different endovascular treatment modalities.

We report a case of female neonate with a progressive choroidal type of VOGM, endovascular treatment of which complicated by thrombosis of the right heart and brain death.

• A 35yo G2P1 pregnant was referred to the Fetal Unite for the 2nd opinion at 33+3 due to dilated blood vessels in the brain. The fetus was found to have a vein of Galen malformation, which had not been identified during the anomaly scan. Ultrasound revealed the mid-line elongated huge cystic structures with the rich arteriovenous flow and mild ventriculomegaly. The fetal MR revealed the impairment of white matter with calcification, leucomalacia and mild posthemorrhagic hydrocephalus.
  • A fetal echocardiogram showed secondary cardiomegaly with the severely dilated right heart and the superior vena cava.
  • No other abnormalities like pericardial and pleural effusion or fetal hydrops were found.

Results Ventriculomegaly progressed rapidly and an elective cesarean section was performed at 36 weeks because of uterine scar and VOGM. The neonate of 2720g had Apgar scores of 6/7 at 1/5 min. Tachycardia up to 240 beats per min, hypotension and seizures developed shortly after birth. MR after birth revealed broadly extended vein Galen with aneurysmal deep and medial superficial vein. Extensive hemorrhagic foci in the white matter were described.

• The total fistula occlusion with Onyx via trans-femoral embolizations were performed on the 3rd day after birth. The procedure complicated by the right heart thrombosis. Multisystem organ failure with brain death was confirmed on the next day after the invasion. The biological death occurred 47 minutes after intensive therapy had been stopped.

Conclusions: In spite of the fact that the outcome of neonates with VGAM has improved tremendously due to advances in endovascular treatment, our case demonstrates that white matter impairment to virtue of venous hypertension and cardiovascular decompensation were the main factors for this outcome. Taking into account the hypoxic-ischemic brain insults and hypertrophic cardiomyopathy as the factors for increased mortality the time of delivery and intervention may be reconsidered.