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Polymicrogyria in a patient after twin-twin-transfusion syndrome

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Introduction

Polymicrogyria (PMG) is a cortical organization, characterized by numerous small irregular and dysplastic convolutions within a gyrus occur[1] Reportedly, 23-26% of cases of refractory epilepsy have underlying MCD. PMG is one of the most common neuronal migration disorders.[2] Twin-twin-transfusion syndrome seems to be revealed as a risk factor for cerebral injuries.[3] Acquired causes of PMG include congenital infections (cytomegalovirus, toxoplasmosis, varicella) and fetal oxygen deprivation secondary to uterine ischemia.[4] Clinical presentation of children with polymicrogyria may include spastic hemiparesis, quadriparesis, developmental delay, dysarthria and severe, treatment refractory epilepsy, which can lead to further neurological impairment depending on the age of first manifestation [5-6] This case report presents a case of PMG after twin-to twin transfusion syndrome with a clinically good outcome.

Clinical presentation & Diagnosis

We report a twenty-six-year-old healthy woman with a monochorionic, diamniotic twin pregnancy in her second pregnancy. During the second trimester the patient developed a twin-twin-transfusion syndrome (TTTS) Quintero stage III and persistent growth discordance of approximately 40%. Selective fetal growth restriction and an oligohydramnios of the donor was observed. The recipient presented with a polyhydramnios, slightly dilated bladder and tricuspid regurgitation. Fetoscopic laser ablation was conducted in gestational week 16+1. The right hemisphere of the donor showed a hyperechogenic area in the parietal lobe. Fetal magnetic resonance imaging was conducted in gestational week 20+3 as seen in figure 3. In the second MRI, blood degeneration producst within the lateral ventricle were found. According to the neuropediatrics prognosis, the findings comply with a risk of 70% for treatment refractory epilepsy. A detailed counseling session was conducted, and the patient decided to continue the pregnancy.

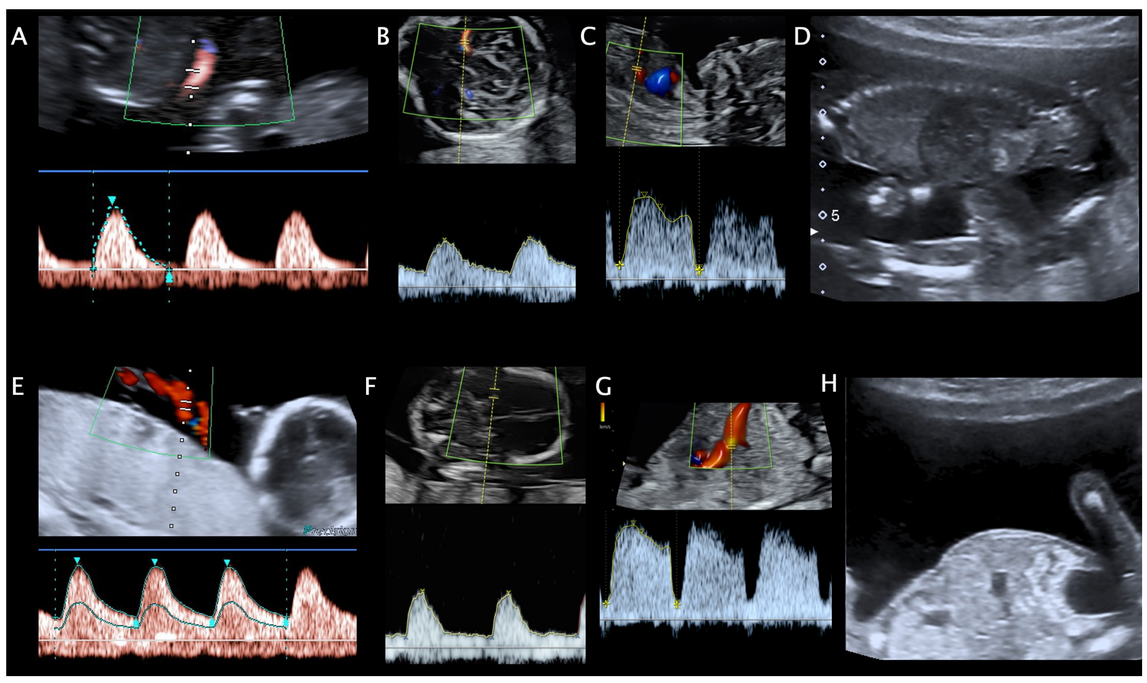

Figure 1. Ultrasound findings at week 15+1 before fetoscopic laser ablation, A)-D) Donor, E)-H) Recipient; A) UA-PI: 2.56, EDF positive B) MCA-PI: 1.75, PSV: 16.1 cm/s C) DV-PIV: 1.22 D) Oligohydramnios, deepest pocket: 1.5cm E) UA-PI: 1.37, EDF negative F) MCA-PI: 1.19, PSV: 23 cm/s G) DV-PIV: 1.11 H) Polyhydramnios, deepest pocket: 7.2cm;

*PI = pulsatility index; UA = Umbilical artery; EDF = End-diastolic flow; MCA = middle cerebral artery; PSV = middle cerebral artery-peak systolic flow velocity; DV = Ductus venosus, PIV = pulsatility index velocity

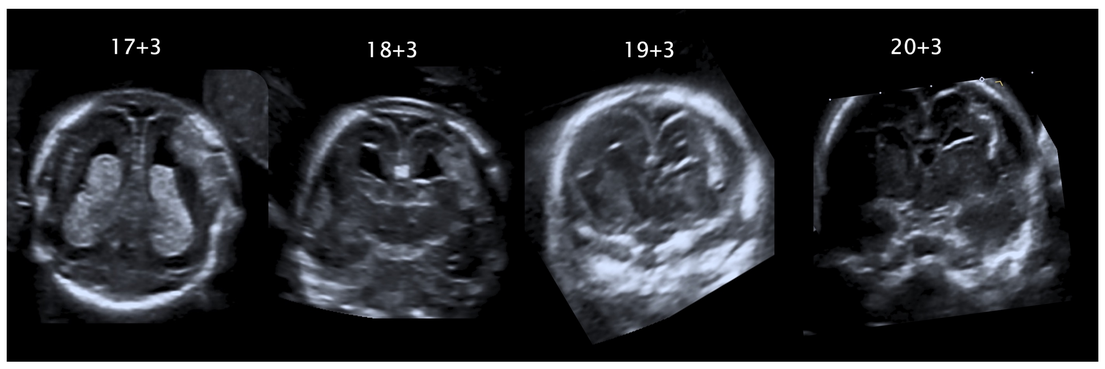

Figure 2. Sonographic presentation of the cortical defect from week 17+3 until 20+3.

Outcome and Follow up

An elective cesarean section was conducted in gestational age of 36+3 weeks without adverse events. Both neonates presented with good APGAR Scores (9/10/10) and normal values of pH and base excess.
Postnatal ultrasound and MRI of the donor verified the prenatally diagnosed defect formation of the parietal right lobe with polymicrogyria and showed the pyramidal tract without myelination. Electroencephalography (EEG) indicated increased seizure propensity, by recording isolated sharpwaves. Clinically, slightly delayed motor development was observed due to a mild left hemiparesis with side preference to the right side. Physical and occupational therapy was provided weekly. Main problems were in the symmetry of motoric function, caused by the impairment of the left upper and lower extremity. Motoric milestones of development depending on the symmetry were delayed by 3-5 months compared to the healthy twin. Free sitting was possible at 13 months, free walking at 19 months. Language and cognitive development did not differ from that of the healthy twin.

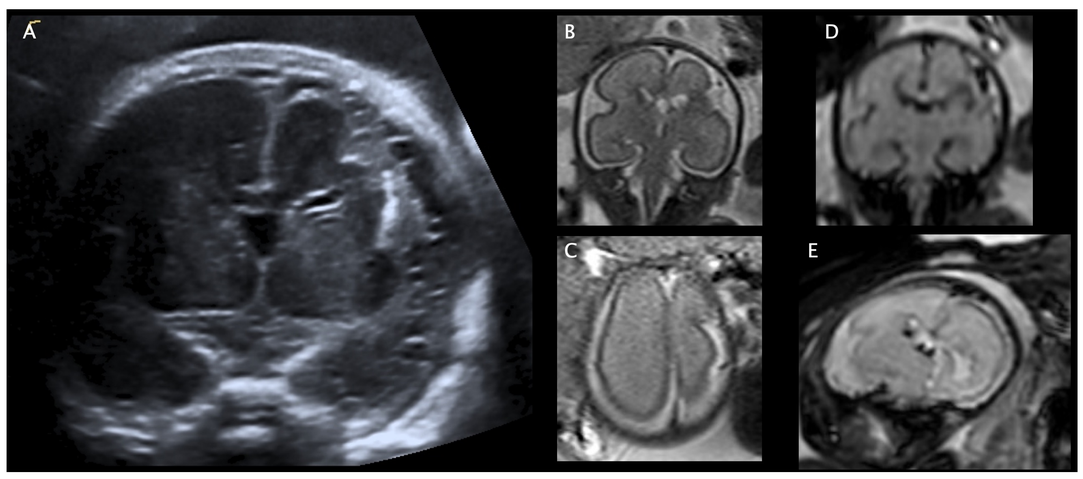

Figure 3. Follow up week 25+3; A) sonography compared to different sections and sequences of the MRI B)-E)

Conclusion

This case report presents a patient with monochorionic twin pregnancy, development of twin-twin-transfusion-syndrome (TTTS) and Polymicrogyria (PMG) of one fetus. Due to TTTS grade 3, fetoscopic laser ablation was performed at gestational week 16+1. After laser ablation, in one fetus, defect formation with PMG of the right parietal lobe was detected. The newborn developed very well, presenting with mild hemiparesis of the left extremities and absence of epileptic seizures despite the expected prognosis.

References

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