Case Report

29-year-old primigravida with Alagille Syndrome (AGS), diagnosed by genetic testing in childhood, presented at 7-weeks by artificial reproductive technology. Preimplantation genetic diagnosis was used to choose an unaffected embryo. AGS is a multisystem autosomal dominant condition caused by heterozygous mutations in the *JAG1* gene, implicated in cell differentiation. AGS is distinguished by five major findings: chronic cholestasis due to intrahepatic bile duct paucity, cardiac abnormalities, posterior embryotoxon, butterfly-like vertebral arch defect, and characteristic facies: prominent forehead, deep-set eyes, mild hypertelorism, straight nose, and small pointed chin.

Due to severe cholestasis, this patient underwent liver transplantation and Roux-en-Y procedure. Her pregnancy was co-managed by MFM and Hepatology. Monthly laboratory tests ensured normal liver function and therapeutic tacrolimus levels. Aspirin prophylaxis was started at 10-weeks. Detailed anatomy ultrasound at 20-weeks excluded cardiac and skeletal anomalies. At 23-weeks, she complained of upper extremity pruritus, that improved on hydroxyzine. At 30-weeks, the fetus was diagnosed with growth restriction. Umbilical artery Doppler surveillance remained normal.

Anesthesiology consultation was requested at 32-weeks given common vertebral defects of AGS. Neuraxial anesthesia was not contraindicated. She presented to labor and delivery at 36-weeks with PPROM and elevated blood pressures, meeting criteria for preeclampsia with severe features. Vaginal delivery was not contraindicated, but she underwent uncomplicated primary cesarean delivery due to malpresentation. Post-operative pain was managed with oxycodone and acetaminophen due to contraindication to NSAIDs. Given the low incidence of Alagille Syndrome (1 in 30,000), this case provides management guidelines and highlights risks of pregnancy with AGS.