

Kaposiform hemangioendothelioma with Kasabach–Merritt phenomenon: Prenatal imaging features & first experience with maternal sirolimus therapy

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Objective

- To describe prenatal diagnosis and perinatal management of **kaposiform hemangioendothelioma (KHE)** complicated by **Kasabach–Merritt phenomenon (KMP)**
- To report the first documented use of **maternal sirolimus therapy (MST)** in this condition

Methods

Retrospective review of 4 fetuses with a prenatal soft-tissue mass suspected of KHE over 18 years-period in a national referring center for superficial vascular anomalies

Results

Prenatal imaging features of KHE

- Marked infiltrative soft-tissue mass
- Doppler : marked hypervascularity
- MRI : T2-hyperintensity

Fetal signs suggesting evolving KMP

- Signs of cardiac decompensation
- Signs of anemia
- Rapid tumor enlargement

MST :

- Initiated in **one severe case** at 34 WG
- Protocol: 1/d, 8 mg/day then 10 mg/day to achieve target trough levels
- Maternal monitoring : lipid panel, liver function tests, fasting glucose, blood pressure, and clinical assessment for mucositis and infection
- **No adverse effect**
- **Prenatal reduction** of infiltration at one month F/U
- **Effective transplacental transfer**
- → Excellent neonatal adaptation, extensive KHE, KMP with severe thrombocytopenia (30 G/L) and markedly elevated D-dimer levels
- → **Postnatal progressive hematologic improvement within 3 weeks**

Conclusion

- ✓ Prenatal imaging can strongly suggest KHE before birth
- ✓ Prenatal imaging may indicate evolving KMP : rapid growth + fetal hemodynamic compromise
- ✓ MST appears feasible and associated with early radiological improvement in 1 severe fetal case



Figure 1. a-d : Axial and coronal T2-weighted fetal MRI at 32 WG : a large submandibular hyperintense infiltrative lesion involving subcutaneous and deep soft tissues. f : Follow-up axial MRI 3 weeks after initiation of MST demonstrates decreased extent of infiltration and reduced T2-signal intensity. g,h: Clinical examination at birth confirmed right lower-face and thoracic wall infiltration, with violaceous skin discoloration

Want to learn more ?

See Fraissenon A et al. Prenatal Diagnosis and Management of Kaposiform Hemangioendothelioma With Kasabach-Merritt Phenomenon: Imaging Features and First Experience With Maternal Sirolimus Therapy. Prenat Diagn. 2026;46(8):1278-1285. doi: 10.1002/pd.70180. Epub 2026 May 16. PMID: 42142389; PMCID: PMC13377337.