

Background

- Fetal cystic lymphangioma is a rare congenital lymphatic malformation characterized by a multiseptated, multicystic lesion that can arise in any organ containing lymphatic vessels
- Existing literature is limited to case reports and case series that often report heterogenous findings, with cases of cystic hygroma and lymphangioma combined

Objective

- Evaluate pregnancy and neonatal outcomes with fetal cystic lymphangioma to facilitate patient counselling

Methods

- A systematic review was conducted in accordance with PRISMA guidance.
- Medline, Embase, Scopus, Cochrane and PubMed Databases were searched utilising the following key:
 - *Fetal OR prenatal OR antenatal OR neonatal OR infant AND: Lymphangioma Fetal lymphatic malformation*
- Inclusion criteria:
 - Case series and case reports published in English between 2000-2022
 - Antenatal ultrasound diagnosis of lymphangioma
- Exclusion criteria:
 - Cases of cystic hygroma

Results

Sixty-two studies (92 fetuses) were included

General characteristics

- Average GA at diagnosis 25.4 weeks
- Commonest locations:
 - 1) axillary 28% (26/92)
 - 2) anterior neck 26% (24/92)
 - 3) abdominal cavity 15% (14/92)

Imaging modality

- All patients were diagnosed via 2D ultrasound
- Fetal MRI done in 44% (40/92)
 - Additional information gained in 85% (34/40)

Associated complications (Figure 1)

- Chromosomal anomalies were reported in 4% (2/52) fetuses
- 1 fetus was diagnosed with Proteus syndrome postnatally

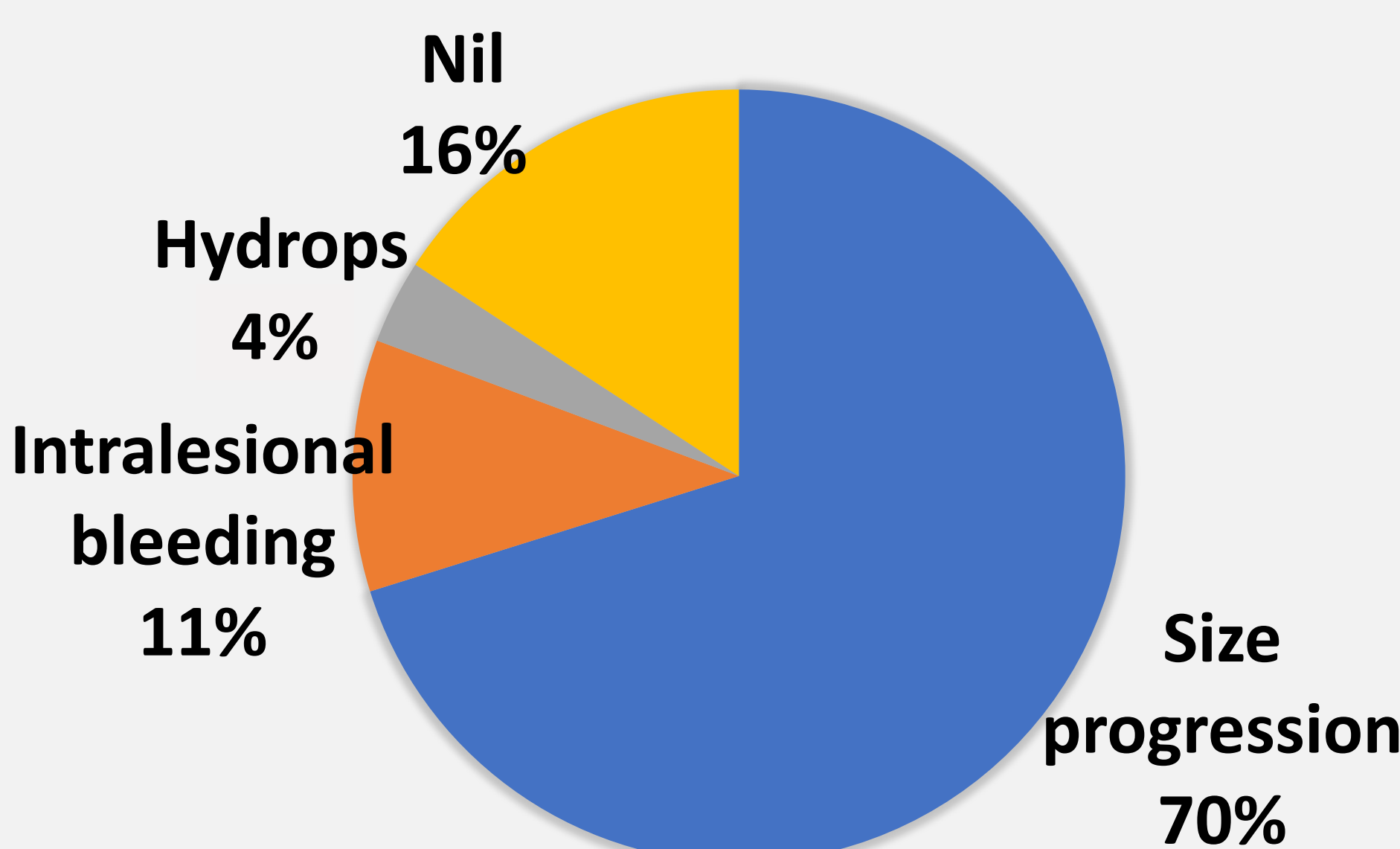


Figure 1: Associated complications

Neonatal outcomes

- Follow-up reported in 91% (68/75)
- Average duration 12.7 months
 - 3 died within one year
 - Remaining 65 had normal development

Pregnancy outcomes (Figure 2)

- Termination of pregnancy 13% (12/92)
- In-utero death 6% (5/80)
- Livebirth rate 94% (75/80)
 - Average GA at delivery 37.7 weeks

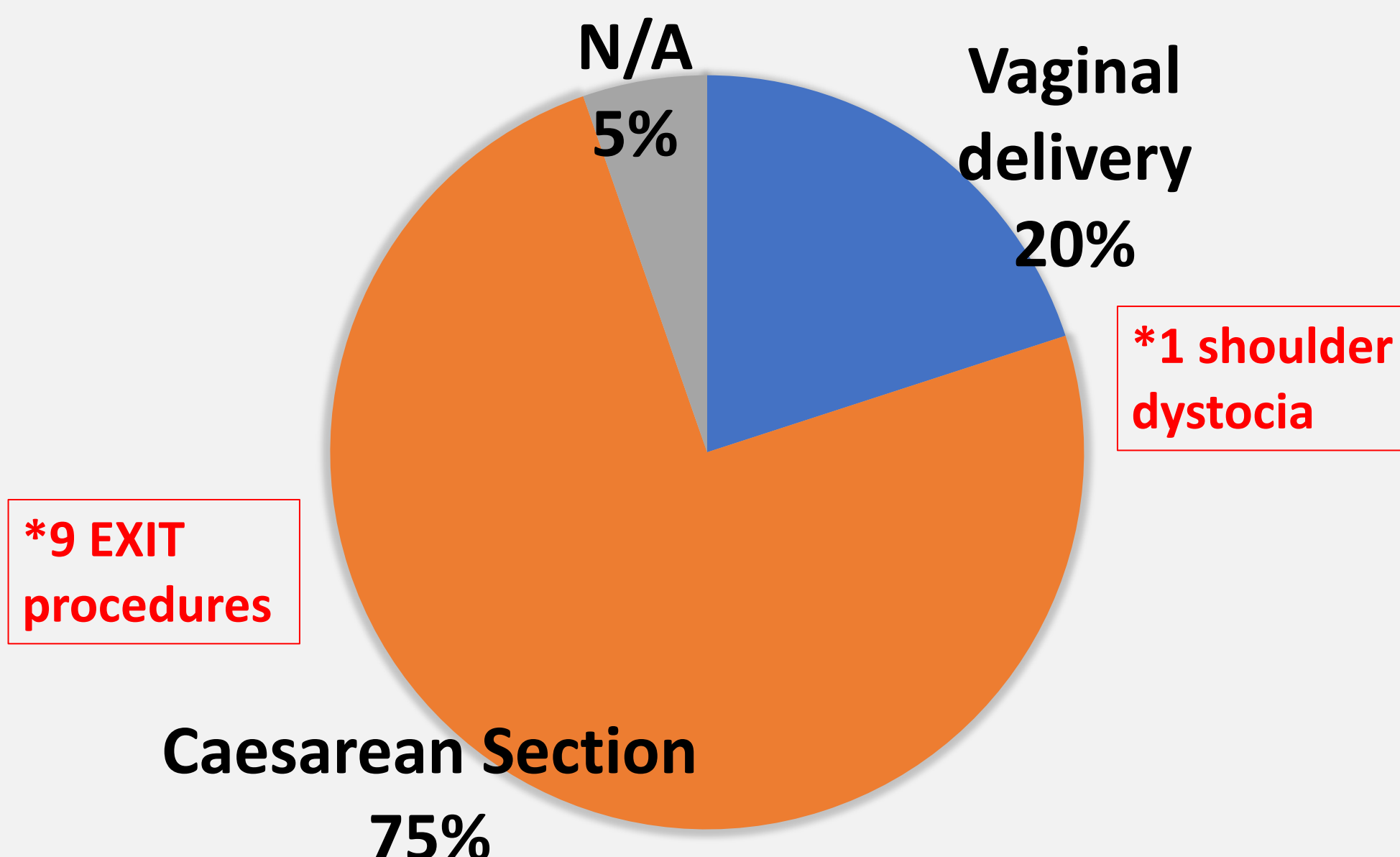


Figure 2: Pregnancy outcomes

Postnatal management

- 1) Surgical excision 39% (29/75)
- 2) Sclerotherapy 20% (15/75)
- 3) Combination of both 12% (9/75)
- 4) Sirolimus or cyst aspiration 6% (5/75)
- 5) Expectant/not reported 23% (17/75)

Conclusion

Fetal cystic lymphangiomas are:

- often detected in the 2nd trimester
- not commonly associated with chromosomal/structural abnormalities
- expected to increase in size during pregnancy
- less commonly associated with Intralesional bleeding and hydrops
- associated with a good developmental outcome

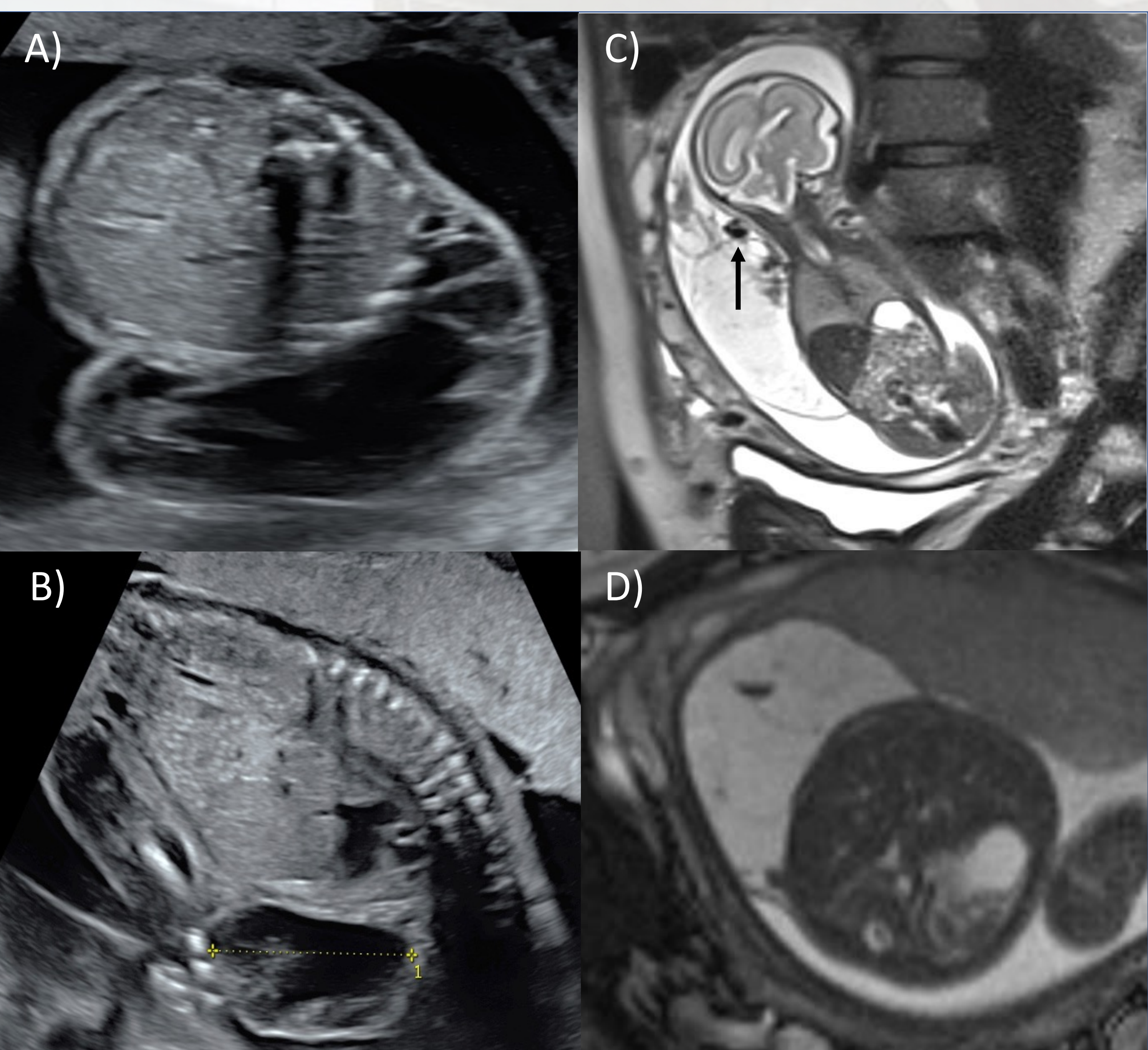


Figure 3: USS (A,B) and MRI (C,D) appearances of fetal lymphangioma presenting with macrocystic lesions and thin septae. Note the blood breakdown products in the cranial part of the lesion (black arrow). A) Sagittal and B) axial US images. C) T2-weighted sagittal and D) axial MRI images.