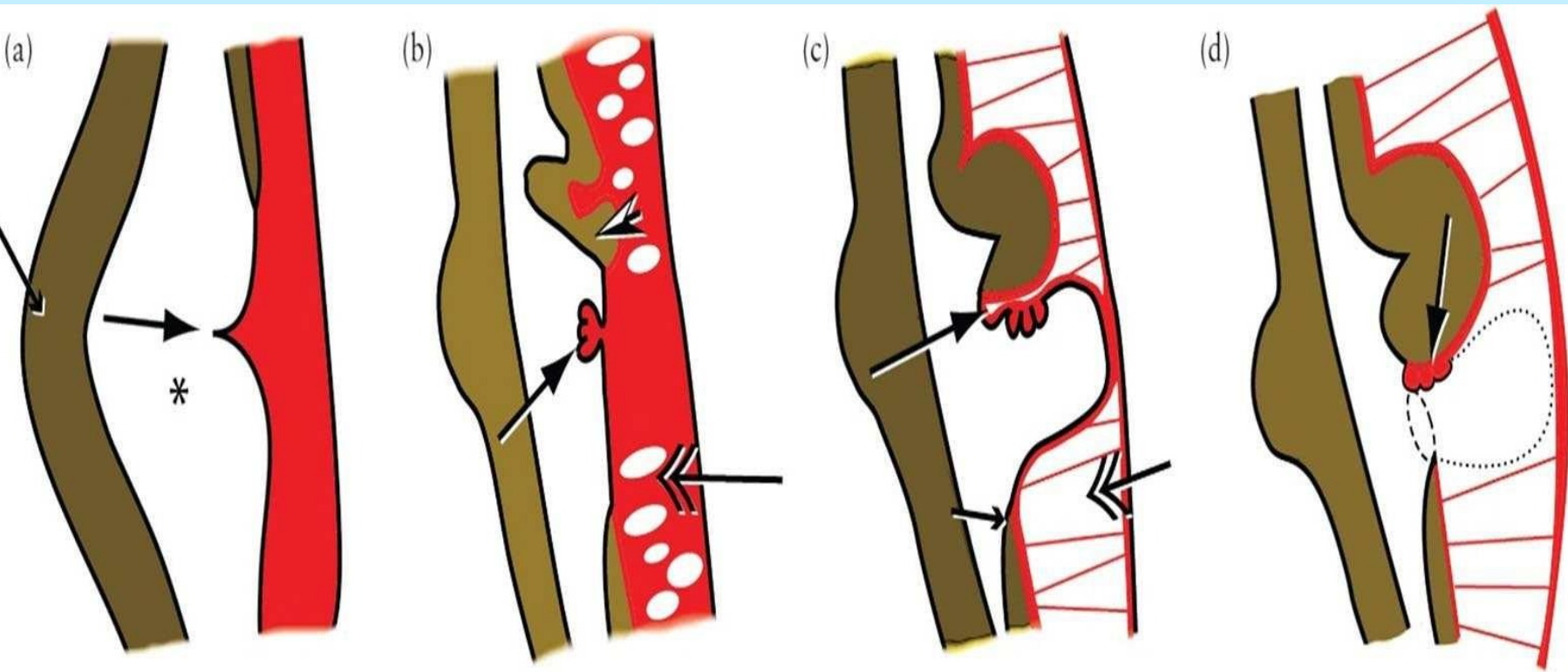
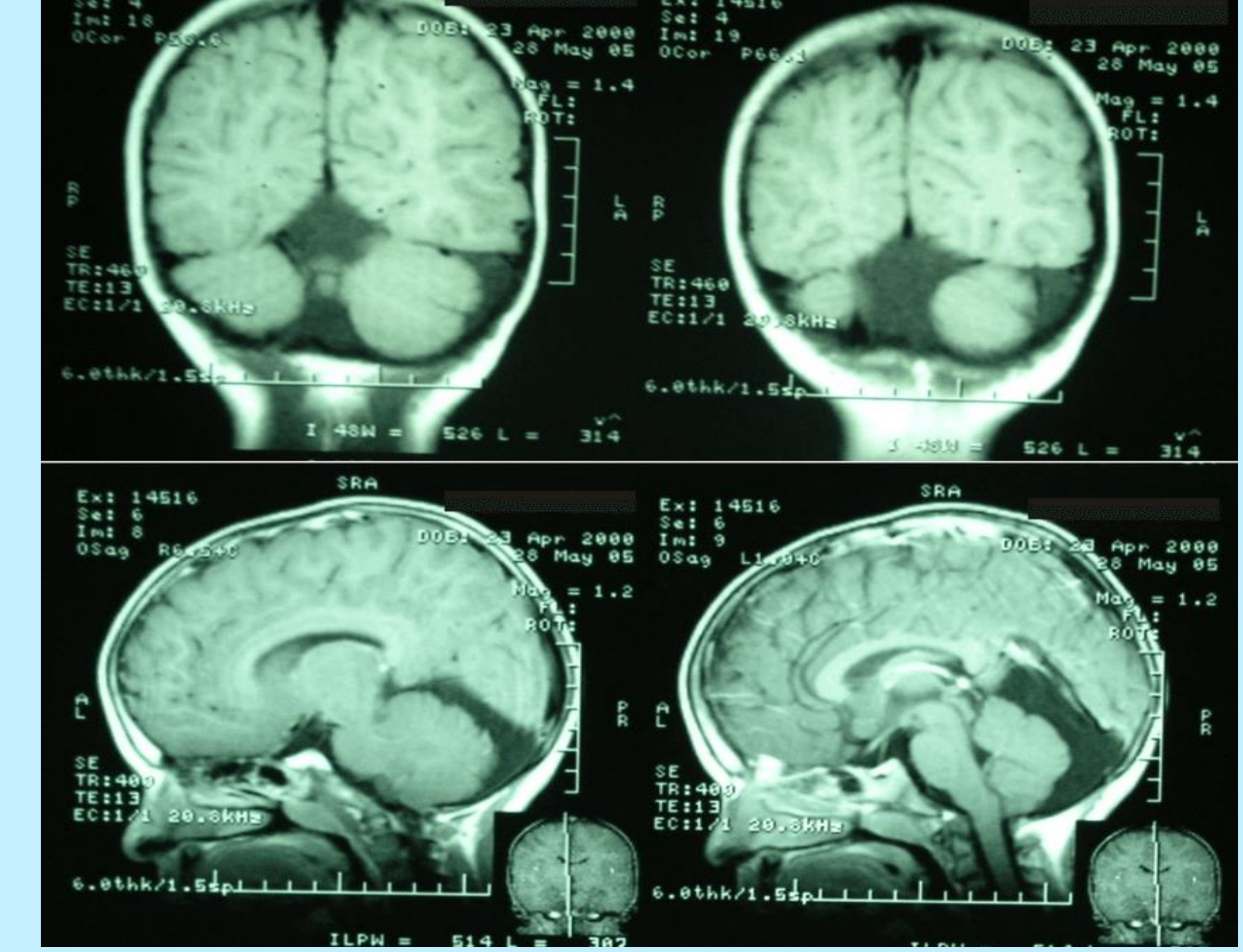


Background :
Isolated inferior vermian hypoplasia (iiVH), also referred as part of Dandy-Walker variant (DWV), is a congenital malformation characterized by partial absence of the inferior portion of the cerebellar vermis. Isolated inferior vermian hypoplasia (iiVH) is one of the most common fetal cerebellar anomalies detected by obstetrical ultrasound and presenting for prenatal counselling.

Objective : We present a case of Vermian hypoplasia detected in the 3rd trimester on ultrasound. The focus of presentation involves the challenges in counseling parents regarding the prognosis and neurodevelopmental outcomes as wide variation exists in the published literature.

Clinical case: A 24 year old female, seen in the outpatient clinic first visit to our hospital, previous antenatal care elsewhere, presented at 31 weeks gestation, found to have SGA on scan and referred for a detailed fetal medicine evaluation. Baby was found to have microcephaly with cerebellar hypoplasia with absent vermis. H/o consanguineous marriage, previous one normal baby and one termination due to fetal anomaly details not available. The couple declined invasive testing and hence further information can only be obtained after birth.

Fig 1. Brain MRI: inferior vermian hypoplasia; asymmetry of cerebellar hemispheres, with hypoplasia of the left cerebellar hemisphere comparative to the right one.



Inferior Vermian Hypoplasia – Pearls and pitfalls

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Literature review:

Isolated inferior vermian hypoplasia (iiVH) is one of the most common fetal cerebellar anomalies detected by obstetrical ultrasound and presenting for prenatal counselling.¹⁻³ Despite its prevalence, the long-term neurodevelopmental consequences of iiVH remain poorly defined and controversial.^{4,5} This is in part due to the Difficulties in reliably diagnosing iiVH, especially by antenatal ultrasound, which provides relatively poor visibility of the posterior fossa structures.⁶ Moreover, the MRI diagnosis of iiVH has a high false-positive rate. (carries a high false positive rate of around 30% antenatally).

There is limited population-based prevalence data for cerebellar malformations, due to several factors. Estimated prevalence varies considerably among available studies(1/3000 to 1/30,000).⁶ Thus, the true prevalence for cerebellar malformations collectively, or for specific disorders, is mostly unknown. Prenatal ultrasound is now widely used to evaluate the fetal brain, and many cerebellar malformations are recognizable before birth. Additional evaluation during pregnancy involves Invasive testing to rule out chromosomal anomalies and fetal MRI.

Fig 2.Postnatal MRI - iiVH



Management :

Includes counseling the parents by the multidisciplinary team including the obstetrician, feto-medicine specialist and the pediatric neurologist. A full workup including a postnatal MRI is crucial in confirming a diagnosis at birth. A close follow up by a pediatric neurologist will help in monitoring the learning and developmental needs of the child.^{7,8}

The real challenge lies in determining the degree of neurodevelopmental outcomes as the prognosis can vary from case to case. This remains the main cause of raised parental anxieties despite best care. A diagnosis of iiVH is associated with persistent elevated parental stress despite normal developmental outcomes in these children suggesting the need for ongoing parental support, even the parents of children with a normal neurodevelopmental outcome in the years following the diagnosis suffered from higher stress levels regardless of their socioeconomic status.^{9,10} This suggests that the diagnosis of iiVH itself carries a significant and long-lasting emotional burden on these families despite the fact that their children are developing normally. Ongoing support and anticipatory guidance should be provided for these families prenatally and postnatally.

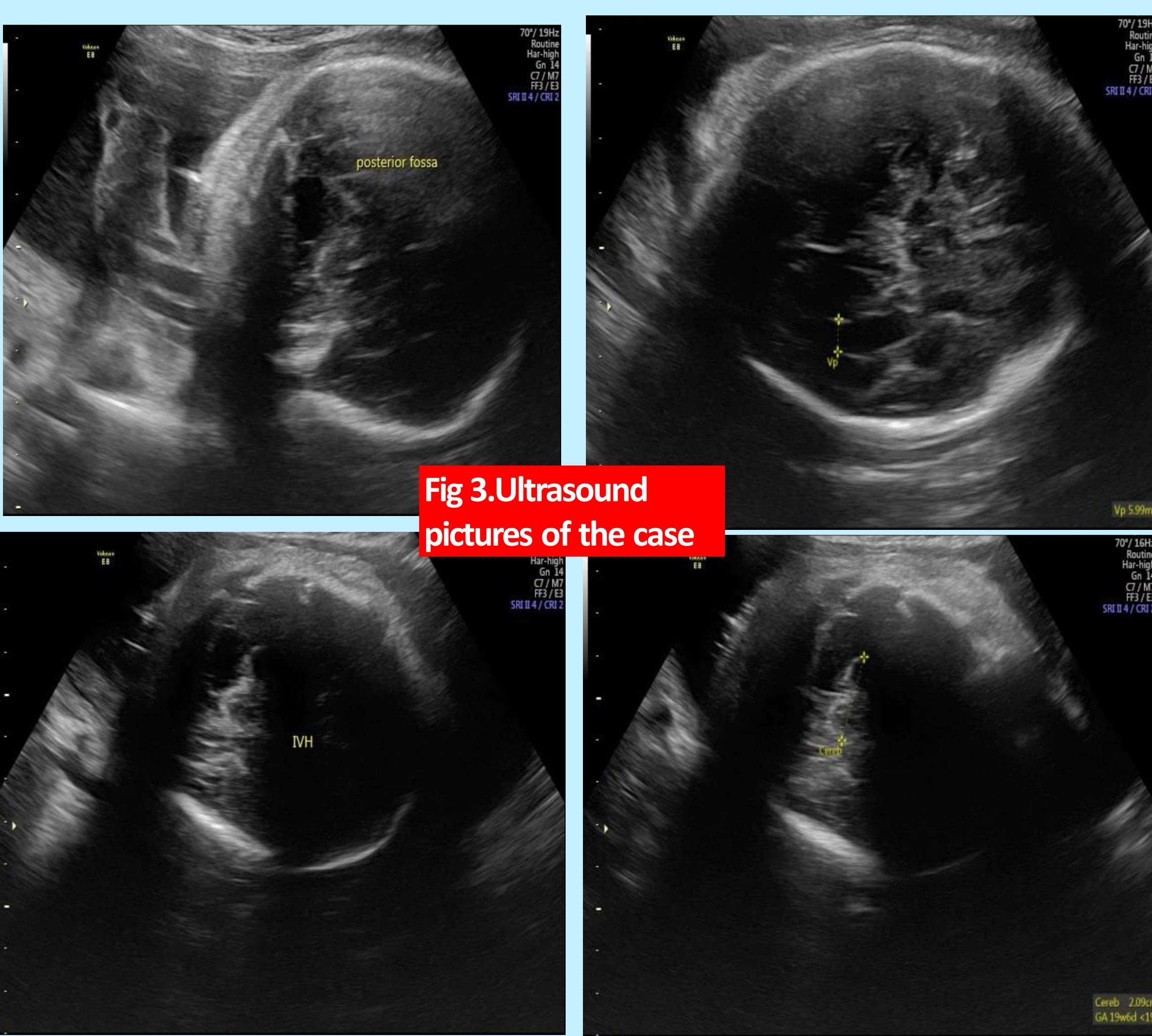


Fig 3.Ultrasound pictures of the case

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