Biallelic pathogenic variants in AEBP1 are the cause of autosomal-recessive EDS subtype. There is only one report of a pregnant EDS patient with the AEBP1 variant, although reports of two female patients among six cases of EDS have been reported. We report the first case of a pregnant Japanese woman who was diagnosed as EDS with a biallelic pathogenic variant in AEBP1 (c.1893C>T, p.Arg631*).

She visited our hospital due to amenorrhea at the age of 34 years. She was diagnosed with EDS at the age of 3 years after presenting with symptoms of motor developmental retardation and joint laxity. She had spontaneous rupture of the transverse colon at the age of 15 years, and a large ovarian tumor (mucinous cystadenoma) was found at the age of 32 years. Following surgery of the transverse colon, she had a post-surgical wound that led to abdominal wall scar hernia. While planning for a baby, we assessed her probable high risk for uterine rupture, digestive tract rupture, and birth canal injury and suggested hospitalization from 20 weeks of gestation, following cesarean section at 23 weeks of gestation. She had ischemia, migraine, and uterine contraction since 23 weeks of gestation. Then, bladder catheterization was performed, and acetaminophen and triptan were prescribed. Magnesium sulfate was intravenously administered for tocolysis. She delivered a 1,947 g female infant by planned cesarean section at 32 weeks of gestation. She had a severe paralytic ileus after cesarean section, which improved with conservative treatment. She was discharged at 22 days after delivery.

![Family tree](Image)

**Medical history**

A 34-year-old Japanese woman (Fig. 1, Fig. 2)

- **Patient**
  - A 34-year-old woman
  - Found to have an AEBP1 homozygous nonsense mutation (c.1893C>T, p.Arg631*).

- **Medical history**
  - 3 years old: Acne
  - 15 years old: Acne
  - 32 years old: Mucinous cystadenoma of the right ovary
  - 33 years old: Biallelic variant of AEBP1

- **Problem lists**
  - Severe pain, paralytic ileus after C/S
  - Acetaminophen and triptan were prescribed.

- **Pregnancy outcome**
  - 6th week: Subchorionic hematoma
  - 21st week: Intermittent lower abdominal pain

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