

## Case Report

# A Sinusoidal FHR Pattern observed in a Case of Congenital Leukemia Diagnosed after Emergent Cesarean Delivery

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### Abstract

Congenital leukemia is a rare disease that develops from birth to six weeks of life and has a poor prognosis. In addition, a prenatal diagnosis is very difficult if risk factors or abnormal echosonographic findings are absent. We herein report a case of congenital leukemia diagnosed after emergent Cesarean delivery due to the presence of a sinusoidal fetal heart rate (S-FHR). At 36 weeks' gestation, the mother was admitted to the hospital because of the loss of fetal movement. An S-FHR pattern was observed almost continuously on cardiotocography (CTG), which was categorized as Category III. A male infant weighing 2,884g was delivered by emergent Cesarean section. The Apgar scores at 1 and 5 min were 3 and 6, respectively, but acidemia was not observed (umbilical arterial pH was 7.259). Because the skin color was pale and general petechiae was apparent at delivery, the baby was transferred to the neonatal intensive care unit and a diagnosis of congenital leukemia was made based on the finding of a thorough examination, including severe anemia (Hb=4.2g/dL, Ht=27.6%) When we observe an S-FHR pattern in patients with undetected risk factors, we should consider the possibility of this clinical entity, despite its extreme rarity.

**Keywords:** Congenital leukemia; Nonreassuring fetal status; Sinusoidal fetal heart rate

### Introduction

Congenital leukemia is a rare condition that is diagnosed from birth to six weeks of life. It occurs at a rate of 1 per 5 million births [1,2]. It has been associated with maternal exposure to radiation, maternal dietary exposure to bioflavonoid, maternal use of tobacco and illicit drugs, and inherited conditions, such as, Down's syndrome, neurofibromatosis, Bloom's syndrome, and Fanconi's anemia [1]. The prenatal diagnosis is very difficult if risk factors or abnormal echosonographic findings such as hydrops due to severe anemia, are absent.

A sinusoidal FHR tracing (S-FHR) is a specific pattern described as a visually apparent, regular smooth sine wave-like undulating tracing seen within the FHR baseline [3]. It is generally thought to be due to severe fetal anemia as well as severe hypoxia.

The continuous S-FHR may appear in situations of severe fetal compromise wherein the fetus has lost total autonomic control. Therefore, the prompt delivery is warranted.

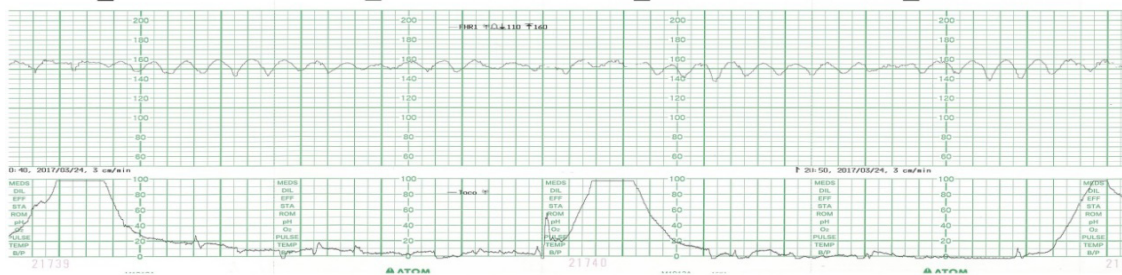
We herein report a very rare case of congenital leukemia diagnosed after emergent Cesarean delivery due to the presence of S-FHR without any risk factors of congenital leukemia.

### Case Report

A 40-year-old Japanese woman (gravida, 0; para, 0) became pregnant without any infertile therapies, and her pregnant course had been uneventful. A non-invasive prenatal test (NIPT) at 12weeks' gestation was negative. The screening tests of fetal ultrasonography were normal at 30 weeks' gestation. None of the above-mentioned risk factors was present.

She was admitted to our hospital at 36 weeks and 6 days' gestation because of the loss of fetal movements. On admission cardiotocography (CTG), an S-FHR pattern was observed (Figure

1). Quick ultrasonography revealed no sign of placental abruption. Fetal hydrops was not confirmed because of the emergent situation. We continued to monitor her for about 1 h, and the finding of an S-FHR pattern was almost continuous, indicating a Category III condition [3]. We discussed the benefits of imminent delivery and performed an emergent Cesarean section due to an unfavorable cervix situation.



**Figure 1:** A Sinusoidal FHR pattern on admission observed in this patient.

A male infant weighing 2,884g was born. The Apgar scores at 1 and 5 min were 3 and 6, respectively, but the acidemia was not observed (umbilical arterial pH was 7.259). Because the skin color was pale and general petechiae were apparent at delivery, the baby was transferred to the tertiary center, which had a neonatal intensive care unit (NICU). His complete blood count was as follows; white blood cell:  $859.09 \times 10^3/\mu\text{L}$  (Blasts 98.5%), hemoglobin: 4.2g/dL, and hematocrit: 27.6%. Because the blast cells were detected on a blood analysis, he was diagnosed with congenital leukemia.

Intensive care such as exchange transfusion, hemodialysis, glucose-insulin therapy for hyperkalemia and administration of rasburicase for hyperuricemia caused by tumor lysis syndrome, were needed to correct his condition. Several B cell makers were positive among blood cell surface markers, and he was diagnosed with B progenitor acute lymphoblastic leukemia. In chromosome tests,  $t(11:19)(q23;p13.3)$ , the MLL-ENL chimera gene, was detected. The maternal postoperative course was uneventful, and she was discharged from our hospital on the postoperative day 7.

## Discussion

As mentioned above, congenital leukemia is a very rare disease, and the possibility of a prenatal diagnosis can be entirely up to chance. Robertson et al. reported that the diagnosis was obtained based on the presence of hydrops on a detailed real-time ultrasound examination at 25 weeks' gestation and it was associated with a Down syndrome revealed by rapid karyotyping [4]. However, the present case lacked any such clues for the diagnosis of this disease [5].

An S-FHR is well known to be due to severe fetal anemia as well as fetal hypoxia. In the past, the term, 'pseudo sinusoidal' has been used but now is not recognized by the National Institute of Child Health and Disease (NICHD) [3]. This term was used to denote a tracing that frequently occurred following narcotic administration or sometimes during ultrasound and was associated with rhythmic fetal movements such as rapid breathing, sucking

movements of the mouth, and thumb sucking, but was not related to fetal hypoxia or anemia [6]. In the present case, 'pseudo sinusoidal' FHR pattern, and fetal hypoxia were not observed. To our knowledge, this is the first report of congenital leukemia showing an S-FHR, although it has been listed as a different diagnosis for the cases of S-FHR.

Determining the relationship between the presence of an S-FHR and the degree of fetal anemia would be extremely interesting. Murata et al. reported that S-FHR was observed in animal experimental models when the hematocrit of fetal lambs decreased to  $<20\%$  [7]. In the present case, fetal hematocrit was confirmed as the value of Ht 27.6% (Hb 4.2g/dL). Because the anemia is not always accompanied in the cases of congenital leukemia [1], we think that S-FHR in this case is caused by fetal anemia, but not specific for congenital leukemia. Of course, it is essential to gather the FHR patterns in the cases of congenital leukemia with and without having anemia, although it might be very difficult because of its extreme rarity.

In cases of chronic onset with hepatosplenomegaly, fetal edema, or polyhydramnios recognized by fetal ultrasonography, if fetal anemia is suspected based on a high middle cerebral artery-peak systolic velocity (MCA-PSV) and the appearance of S-FHR, congenital leukemia may be considered as a differential diagnosis. However, in acute-onset cases, such as the present case, a pre-delivery diagnosis may be impossible.

Physicians should consider a possible diagnosis of congenital leukemia when S-FHR appears, even in cases without any of the traditional risk factors. In addition, congenital leukemia may be involved in cases of unknown intrauterine fetal death, so the further accumulation of similar case reports is warranted.

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