

## CASE REPORT

# Hypoplasia of abdominal wall muscles following massive fetal persistent chylous ascites without anemia

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## Abstract

Abdominal wall hypoplasia is a widely known clinical finding of genetic disorders such as the prune belly syndrome. On the other hand, there are few cases of abdominal wall muscle hypoplasia associated with fetal ascites due to fetal hydrops caused by fetal anemia have been reported. We report a case of fetal chylous ascites without anemia, resulting in abdominal wall muscle hypoplasia and flabby skin. At 17 weeks of gestation, fetal ascites was first detected and deteriorated without anemia. At 28, 33 and 36 weeks of gestation, paracentesis was performed three times because of cardiovascular impairment, confirming chylous ascites. After birth, the baby exhibited a flabby skin and lateral abdominal wall hypoplasia, resulting in difficulties in maintaining a sitting posture at 10 months of age. The genetic test using the TruSight One Sequencing Panels found no genetic variants. This case suggests that abdominal wall hypoplasia could be associated with fetal ascites without anemia.

## INTRODUCTION

Abdominal wall muscle hypoplasia associated with fetal ascites due to anemia in utero has been reported in several cases [1–3]. Although tissue hypoxia has been suggested to may be caused by fetal anemia in those cases, the precise etiology remains unclear. Herein, we report a case that manifested an isolated abdominal wall muscle hypoplasia and a remarkable flabby skin following massive fetal persistent chylous ascites without anemia *in utero*. He did not harbor known mutations as a cause of a genetic disease, which resulted in difficulties of keeping a sitting posture until 10 months of age. This case demonstrates the possibility that fetal persistent ascites without anemia in

*utero* might be related to the congenital abdominal wall muscle hypoplasia.

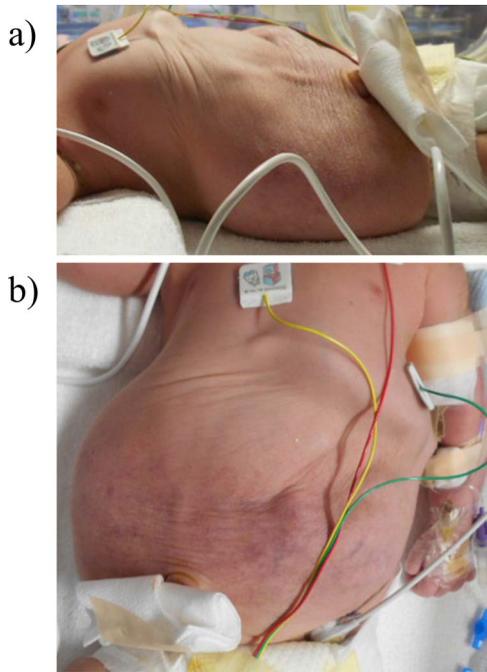
## CASE REPORT

A 39-year-old woman, gravida 2, para 1, was referred to our hospital at 13 weeks of gestation because of positive anti-M antibody at the serological screening test. Although no abnormal finding was found on ultrasound examination (US) at the initial visit, fetal left hydrothorax was detected at 15 weeks and ascites at 17 weeks. Maternal serological tests for parvovirus B19 (PB19), cytomegalovirus, toxoplasmosis, rubella and syphilis were nega-

Received: July 20, 2020; Revised: January 9, 2021; Accepted: March 27, 2021

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**Figure 1:** Physical examination of the newborn at birth, showing abdominal skin wrinkles (a) and bilateral abdominal wall projection (b).

tive, and amniocentesis confirmed a normal karyotype denoted as 46 XY. Although fetal hydrothorax spontaneously regressed from 19 weeks and completely disappeared at 21 weeks, fetal ascites deteriorated, and fetal echo revealed holosystolic tricuspid valve regurgitation at 21 weeks. At 27 weeks of gestation, reverse flow in the ductus venosus and increased preload index in the inferior vena cava without an elevated peak systolic velocity of the middle cerebral artery occurred, suggesting cardiovascular impairment. At 28 weeks of gestation, paracentesis was performed to improve the fetal cardiovascular impairment, and fetal ascitic fluid contained 92% lymphocytes, deducing chylous origin. A second paracentesis was also performed at 33 weeks for the same reason. However, fetal ascites increased again, and maternal abdominal distension with polyhydramnios worsened. Therefore, elective cesarean section was performed due to previous cesarean section at 36 0/7 weeks of gestation along with the third paracentesis in order for the baby to be delivered easier.

A male infant weighing 3488 g with a symmetrically enlarged lateral abdomen associated with abdominal skin folds and cryptorchidism was delivered (Fig. 1). Total parenteral nutrition was introduced, and intravenous administration of octreotide was initiated since Day 6, followed by a low-fat formula diet containing medium-chain triglycerides (MCT) from Day 15. Octreotide was terminated on Day 36, and breast feeding was started on Day 59. US and computed tomography showed worsened ascites after increasing breast feeding; therefore, breast feeding was switched to low-fat formula containing MCT diet on Day 77. The ascites decreased, and the baby was discharged on Day 92. The baby started rehabilitation to improve his abdominal muscle strength and movement. At 10 months of age, he had difficulty keeping a sitting posture, although neurological findings were normal. At 11 months of age, magnetic resonance imaging showed an oblique and transverse abdominal muscle hypoplasia (Fig. 2). Next, orchiopexy was



**Figure 2:** T1-weighted magnetic resonance image (axial view) of the abdomen obtained at 11 months of age, below the umbilical level. Oblique and transverse abdominal muscles at both sides are thin (1, 2), especially on the right side (1). In contrast, the rectus abdominis muscle is thin to a lesser degree (3).

performed at 12 months of age, and we confirmed that his oblique and transverse muscles are very thin and vulnerable. Genetic blood test was performed using TruSight One Sequencing Panels (Illumina Inc., San Diego, CA), which covered ~4800 disease-associated genes, and no genetic variants were found. Although he exhibited normal fine and gross motor functions without any neurodevelopmental disabilities at 19 months of age, cosmetic problems of abdominal distension and flabby skin persisted.

## DISCUSSION

Abdominal muscle hypoplasia following the fetal ascites is previously reported. Embryological studies suggest that diastasis recti abdominis would occur due to disruptive phenomena before the fusion of myotomes during the 8 weeks of gestation, whereas muscular hypoplasia should arise from the later phenomena [1]. Only four case reports of abdominal muscle hypoplasia associated with fetal ascites have been published in English language [1–3]. One of them had ascites from 22 weeks due to twin-to-twin transfusion syndrome (TTTS), whereas the others manifested ascites from 20 to 22 weeks due to PB19 infection. All of them showed lateral abdominal bulging at birth, having hypoplastic oblique and transverse abdominal muscles with normal recti abdominis. Travan et al. supposed that muscle hypoplasia is not only attributed to mechanical compression with vascular compromise inducing ischemia due to fetal ascites but also to fetal anemia caused by TTTS and PB19 infections, which provoke tissue hypoxia [3]. Note that our case had chylous ascites from 17 weeks without anemia; therefore, its pathogenesis is different from the four previously reported cases. Several genetic disorders, such as prune belly syndrome (PBS), are supposedly caused by abdominal muscle defects, and PBS is characterized by laxity of the abdominal wall musculature, bilateral cryptorchidism and urinary tract anomalies [4]. Our case did not present uropathy during the prenatal and postnatal periods, suggesting it was different from PBS.

Congenital chylothorax and chylous ascites could occur during fetal life due to lymph leakage into the pleural and/or peritoneal cavities, and their causes include inappropriate chyle transport toward the main circulation because of aplasia, hypoplasia, obstruction or thoracic duct severance; they are idiopathic in numerous cases [5]. Moreover, spontaneous regression of primary fetal hydrothorax commonly occurs in

9–22% of cases [6], whereas only one report described a case of fetal chylous ascites that spontaneously regressed before birth [7]. In this case, cytological examination of the fetal peritoneal fluid revealed chylous ascites. Therefore, the hydrothorax might be chylothorax; however, although chylothorax regressed during pregnancy, fetal chylous ascites occurred and persisted throughout pregnancy. To our knowledge, this is the first case report on such a case, although the etiology remains unclear.

In conclusion, our case demonstrates that persistent fetal ascites from the early second trimester without anemia could be associated with congenital abdominal wall muscle hypoplasia.

## CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

## FUNDING

There is no source of funding.

## ACKNOWLEDGEMENTS

None declared.

## ETHICAL APPROVAL

Not applicable.

## CONSENT

Informed consent was obtained from the patient for publication of this case report and accompanying images.

## GUARANTOR

Shunsuke Tamaru.

## REFERENCES

1. Macé G, Audry G, Cortey A, Nguyen A, Slaim L, Castaigne V, et al. Congenital hypoplasia of the abdominal wall muscles following fetal ascites due to parvovirus B19 infection. *Ultrasound Obstet Gynecol* 2011;**37**:497–8. doi: [10.1002/uog.8904](https://doi.org/10.1002/uog.8904).
2. Lubinsky M, Rapoport P. Transient fetal hydrops and “prune belly” in one identical female twin. *N Engl J Med* 1983;**308**:256–7. doi: [10.1056/NEJM198302033080505](https://doi.org/10.1056/NEJM198302033080505).
3. Travan L, Naviglio S, Cont G, Brovedani P, Davanzo R, Demarini S. Isolated hypoplasia of abdominal wall muscles associated with fetal ascites. *Congenit Anom (Kyoto)* 2016;**56**:184–6. doi: [10.1111/cga.12156](https://doi.org/10.1111/cga.12156).
4. Shimada K, Hosokawa S, Tohda A, Matsumoto F, Johnin K. Histology of the fetal prune belly syndrome with reference to the efficacy of prenatal decompression. *Int J Urol* 2000;**7**:161–6. doi: [10.1046/j.1442-2042.2000.00159.x](https://doi.org/10.1046/j.1442-2042.2000.00159.x).
5. Lopez-Gutierrez JC, Tovar JA. Chylothorax and chylous ascites: management and pitfalls. *Semin Pediatr Surg* 2014;**23**:298–302. doi: [10.1053/j.sempedsurg.2014.09.011](https://doi.org/10.1053/j.sempedsurg.2014.09.011).
6. Trocado V, Coutinho-Borges JP, Carlos-Alves M, Santos J, Pinheiro P. Fetal primary hydrothorax with spontaneous resolution. *Case Rep Womens Health* 2017;**15**:6–7. doi: [10.1016/j.crwh.2017.06.001](https://doi.org/10.1016/j.crwh.2017.06.001).
7. Chéreau E, Lejeune V, Gonzales M, Carbonne B. Voluminous fetal chylous ascites: a case of complete spontaneous prenatal regression. *Fetal Diagn Ther* 2007;**22**:81–4. doi: [10.1159/000097101](https://doi.org/10.1159/000097101).