

Case Report

Report a Case of Congenital Adactyly Associated with Intestinal Obstruction in the Third Baby of a Triplet Pregnancy after *In Vitro* Fertilization

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Abstract

Congenital adactyly/hypodactyly is an extremely rare musculoskeletal defect consisting of a transverse terminal deficiency of digits. In this report a case of congenital absent digits has been introduced that was associated with jejunal atresia in the third baby of a triplet pregnancy who was born by IVF technique.

Keywords: Congenital adactyly; Congenital absent digits; Jejunal atresia; Intestinal obstruction; ART

Introduction

According to reports fertilization after Assisted Reproductive Technology (ART) have spread worldwide and the number of births after ART have been increasing steadily [1,2]. On the other hand, multiple embryos are transferred in most ART techniques contribute to multiple gestation pregnancies in many cases that may lead to risks to the babies including prematurity, low birth weight, death and greater risk for birth defects [3-6].

Congenital absent digits is a rare disorder that defined by many difficult confusing terms such as adactyly, symbrachydactyly, ectrodactyly, amniotic band syndrome [7]. The sporadic form's occurrence rate is 1/10000 live births [8]. An international group of clinicians has introduced the re-definition of all terms to standardization of them and consensus regarding their definition [9]. The categories that they defined were subdivided into non-syndromic and syndromic forms [9].

In this report a case of congenital absent digits has been introduced that was associated with jejunal atresia in the third baby of a triplet pregnancy who was born by IVF technique. To the base of search in literatures this is the first case of congenital adactyly associated with jejunal atresia in a neonate born by ART.

Case Report

Three newborns which were the result of a preterm 29 weeks a triplet pregnancy were admitted in NICU because of respiratory distress syndrome. They conceived by *In Vitro* Fertilization (IVF) after 8 years primary infertility of non-related parents. Third baby was a boy weighing 880 grams and physical examination revealed symmetrical absence of digits in feet and hypodactyly in both hands as well (Figure 1). Other exams and surveys including brain ultrasonography and cardiac echocardiography were normal. No facial dysmorphism was found. Two other babies had no dysmorphic appearance. Respiratory distress managed by mechanical ventilation and surfactant administration. In second day of admission, abdominal distention occurred and neonate developed bilious vomiting. Abdominal plain film showed dilated, gas-filled bowel loops and



Figure 1: Photograph showing hypodactyly in hands and adactyly in feet.

absence of rectal gas (Figure 2). The patient underwent an operative repair on third day of life and post-operative diagnosis was jejunal atresia. After surgery the baby was critically ill and he presented with severe pulmonary hemorrhage on day 10th of life not responding to all medical treatments. Unfortunately, he died due to respiratory failure and massive pulmonary hemorrhage despite of all managements on day 12th of life.

Discussion

In our report a case of congenital hypodactyly/adactyly who conceived by IVF has been reported. Premature birth and congenital birth defects are complex conditions related to infant's mortality and morbidity [2]. The risk of complications and congenital anomalies after assisted reproductive technology is still a controversial and debated subject. Some of the complications have been related to a higher rate of multiple births and increased risk of preterm delivery; however other studies showed that even singleton IVF babies have a higher rate of low birth weight and birth defects [2]. In a recent meta-analysis that assessed the use of ART on the risk of birth complications and congenital anomalies the odd's ratio for preterm labor was 1.79 (95% CI 1.21 to 2.63), low birth weight 1.89 (95% CI 1.36-2.62) and musculoskeletal disorders 1.35 (95% CI 1.12 to 1.64) which were all



Figure 2: Gaseous distention of bowel loops and absent rectal gas in abdominal radiography.

statistically significant. In the present study risk of musculoskeletal disorder in babies conceived by ART was 35% higher than newborns of normal conception [10].

Congenital adactyly/hypodactyly is a musculoskeletal defect consisting of a transverse terminal deficiency of digits. This defect occurs in 3 forms including: sporadic, as a dominantly inherited trait and as a part of syndrome [8]. Hypoglossia-hypodactyly (aglossia-adactyly) syndrome is a very rare disorder characterized by a small tongue associated with limb defects including adactyly, oligodacty or syndactyly [11]. In this case report there was no history of birth defects in family and the newborn's tongue exam was normal. On the other hand, in this presented case report patient conceived by IVF and had intestinal obstruction as well.

Donahue et al reported severe upper limb reduction and duodenal atresia in a premature infant born at 32 weeks gestation who was the product of a normal triplet pregnancy. Although the hands of patient showed two syndactyly digits with bifid nail, the major problem was arm reduction associated with other multiple anomalies. Finally, karyotype analysis showed 6q16.3 micro deletion syndrome [12]. The main difference of above case with Donahue's report is this point that no other anomalies had been detected and patient was an ART-conceived baby.

Even though higher rate of musculoskeletal defects has been reported in babies born after ART, it seems the limb defect in presented case is not merely due to ART mechanisms. The noticeable point is that congenital absent digits may be categorized by many confusing terms. For example many clinicians have difficulty distinguishing between symbrachydactyly, transverse deficiency, constriction ring syndrome or amniotic band syndrome [7]. Previous studies showed that vascular accident is a major etiology of intestinal atresia of the jejunum [13]. On the other side terminal transverse limb defects and amniotic band syndrome have been attributed to the process of

vascular disruption [14]. So the limb defects and intestinal atresia in this presented case may be occurred at the first trimester due to vascular accident as a common cause of the event.

Conclusion

Congenital adactyly is a very rare defect and its exact embryogenesis is enigmatic. In spite of the low incidence of congenital limb defects in the ART-conceived infants, assessment of other complications such as intestinal obstruction as an associated anomaly especially in multiple gestation pregnancy after ART can lead to early management and can be life-saving.

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