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Case Report

Pregnancy in a Previously Conjoined Thoracopagus Twin with a Crisscross Heart

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Background. Crisscross heart (CCH) is a complex, rare, congenital, rotational, cardiac abnormality that accounts for <0.1% of congenital heart defects (CHD). CCH is characterized by the crossing of the inflow streams of the two ventricles due to an abnormal twisting of the heart. A case of maternal CCH has not been previously reported. Case. We report a case of a primigravida with CCH, who was separated at birth from her thoracopagus conjoined twin. Pregnancy was managed by congenital cardiology, maternal-fetal medicine, anesthesiology, and obstetrics. She underwent a 39-week vaginal delivery without maternal or neonatal complication. Conclusion. A successful term pregnancy outcome was achieved in a patient with CCH using a multidisciplinary approach to address her cardiac condition.

1. Introduction

The incidence of multiple gestations continues to increase, now accounting for more than 3% of all live births in the United States [1]. Conjoined twins are the rarest subset of monochorionic gestations, occurring in approximately 1 in 50,000 to 1 in 190,000 births [2]. The majority of conjoined twins do not survive and their outcome depends on the type of fusion and associated defects [2]. Conjoined twins are classified according to the most prominent shared anatomy. Thoracopagus classification accounts for 75% of conjoined twins and describes fetuses that have a common sternum, diaphragm, upper abdominal wall, liver, pericardium, and gastrointestinal tract [2]. Most of the thoracopagus twins share cardiac anatomy and/or a congenital heart defect (CHD) is present in one or both twins [2].

Crisscross heart (CCH) is a complex, rare, congenital, rotational abnormality that accounts for <0.1% of congenital heart defects (CHD) [3]. This CHD is characterized by the crossing of the inflow streams of the two ventricles due to an apparent twisting of the heart about its long axis. In general, neonates born with CCH have an overall poor outcome with cyanosis commonly noted at birth. Depending on the severity of other cardiac anomalies that are commonly associated with CCH, such as transposition of the great arteries, double outlet right ventricle, pulmonary stenosis, and atrioventricular defects, symptoms of heart failure can be also encountered at birth [3]. Pregnancy among patients with CHD is associated with maternal and fetal complications.

A literature search did not reveal any prior cases of pregnancy in women with CCH; we herein present a case of a primigravida with CCH separated at birth from her thoracopagus twin who underwent successful term vaginal delivery without complication.

2. Case Report

A 29-year-old gravida 1, para 0, with a history as a thoracopagus conjoined twin presented to our prenatal clinic at 15 weeks of gestation. She underwent a successful separation from her sister at 10 months of age. Her cotwin has no medical problems with respect to her history as a conjoined twin and specifically no cardiac complications. Conversely, our patient’s cardiac anatomy was complicated by a CCH, situs solitus with dextrocardia, and a secundum-type atrial septal
Conjoined twins are the rarest subset of monozygotic twins occurring as a result of incomplete zygotic division that occurs 13 to 15 days after fertilization. In general, 25%–30% will die prior to delivery, 40%–50% immediately after birth, and 15%–20% surviving thereafter [4]. There are no trials available regarding the most appropriate management of these affected pregnancies and termination is usually recommended. In particular, the presence of thoracopagus conjoined twins as this type is frequently accompanied by shared cardiac anatomy and prognosis for surgical division is very poor [4].

Conjoined twins and CCH are very rare amongst themselves, and the combination of both occurring in a pregnant mother has not been previously described in the literature. Amongst the other described rare congenital heart defects in pregnant women, successful pregnancies have occurred in patients with a single ventricle and transposition of the great arteries [4]. As compared to those case reports, a successful pregnancy was achieved in this case without any major complications.

The presence of the bioprosthetic tricuspid valve in relation to her CCH was of particular concern in our patient. The valve was seated abnormally from its typical position due to the angulation associated with CCH. As such, the gradient across the valve was increased despite the leaflets themselves being of normal function. This along with a right ventricle that was smaller than usual due to her history of Ebstein’s was a significant concern as to how her right ventricle would tolerate the volume load of pregnancy. Fortunately, her right ventricle tolerated the pregnancy well and she only required a few doses of oral diuretics postpartum.
The desire for a patient with a complex congenital heart defect to become and/or continue a pregnancy should be evaluated on a case-by-case basis taking into consideration their baseline functional status and anticipated response to pregnancy changes based on their underlying pathophysiology. The maternal/perinatal risks should be reviewed with these patients using the best information we have. A multidisciplinary team is required in the pregnancy planning and management. This team approach requires active participation of maternal-fetal medicine specialists, adult congenital heart specialists, obstetricians, and anesthesiologists with experience in managing the pregnant women with congenital heart disease.

**Disclosure**

This paper is not under consideration by any other journal.

**Conflict of Interests**

None of the authors declare any conflict of interests in the paper, including financial, consultant, institutional, and other relationships that might lead to bias or a conflict of interests.

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