

Case report:

Omphalopagus Conjoined Twins: A Case Report

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Abstract:

We present a case of conjoined omphalopagus twins in a 35-year-old woman who came for antenatal booking at 9 weeks of gestation to a health centre in Kota Bharu, Kelantan. The diagnosis was confirmed via ultrasonography by an obstetrician in a tertiary Hospital at 12 weeks of gestation. The pregnancy was allowed to progress as it carried the best chance of survival. However, the parents were in dilemma with the decision. Primary care practitioners play an important role to prepare patients mentally and physically to face the outcome of the pregnancy.

Keywords: conjoined twins; omphalopagus twins; Siamese twins; twin pregnancy

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Introduction

Conjoined twins are a rare form of twins gestation and it is associated with high perinatal mortality. Popularly known as Siamese twins, it is a congenital anomaly that results from abnormal embryogenesis. It has been postulated to occur during the zygote formation when there is incomplete cell division between the 13th and 15th days after fertilization.¹ Conjoined twins are classified based upon the site of attachment. It is estimated to occur in approximately one in 250 000 live births.² In Malaysia, conjoined twins occur in one in every 250 000 to 500,000 live births.³ Based on the site of the union, conjoined twins could be ventral in 87% of the cases and dorsal in 13% of the cases. Ventral unions are classified to the thorax (thoracopagus-19%), abdomen (omphalopagus-18%) and pelvis (ischiopagus-11%). Dorsal unions include skull (craniopagus-5%),

sacrum (pygopagus-6%) or back (rachipagus). Most conjoined twins are female in predominance.²

Case Report

A 35-year-old G₅P₃₊₁ presented to a government health clinic in Kota Bharu for booking at 9 weeks period of gestation. She was otherwise asymptomatic and had no underlying medical problems. Her menstrual history was regular, and she was sure of her last menstrual date. Her previous pregnancies were uneventful. There was a strong family history of twins on the paternal side but there were no incidences of conjoined twins before.

A routine physical examination which included thyroid, breast and abdominal examination was done and it was unremarkable. An ultrasound scan was performed by the medical officer to confirm foetal viability, and it showed a grossly abnormal foetus (Figure 1).

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Figure 1: Image showing abnormal fetus at 9 weeks' gestation

She was then referred to a tertiary hospital nearby for a confirmatory diagnosis. A detailed ultrasound was done by an obstetrician at 12 weeks of gestation (Figure 2).



Figure 2: Image of conjoined twins (omphalopagus) at 12 weeks' gestation

The transabdominal ultrasound scan had shown the presence of two heads, four arms and four legs. The area of union was localized to the abdominal region with a shared liver (Figure 3).

There were two umbilical cords and the placenta was localized posteriorly. Amniotic fluid was within normal limits. Thus, the diagnosis of an omphalopagus conjoined twins was confirmed.

The parents were informed in detail about the malformation. The discussion was done on the outcome of that special pregnancy, which bore a good chance of survival for both twins. They were given ample time to think and decide. Even though in the beginning the parents could not accept the information



Figure 3: USG showing fusion of the liver

and were not keen to continue with the pregnancy, they finally opted to let the pregnancy progress.

Discussion

It is reported that among risk factors for foetal anomaly is diabetes mellitus.⁴ By saying that, the patient is free of any risk factors which might not explain why she got conjoint twin pregnancy. Twins pregnancy, in general, might give a different joy to the parent but having conjoined twins may cause another complete surprise. Adequate knowledge about this rare twins pregnancy could help the parents have a better understanding and prepare them in the decision making regarding the pregnancy. The overall survival rate of conjoined twins is from 5-25%.⁵ For better obstetric management and treatment planning, early correct prenatal diagnosis is essential. Early antenatal booking at the primary care clinic allows early routine ultrasound and detection of the conjoined twins. For medical officers in primary care, it is very important to recognize the conjoined twins since early referral is needed to assess the extent of fusion and the functioning organ. There are conjoined twins reported as early as the 10th week of gestation.⁶ This can be done with help from advanced development in imaging modalities.¹ Once the diagnosis is confirmed with ultrasound, the possible outcome of the pregnancy can be predicted

from the characterization of the type and severity of the abnormality seen by the MRI. Conjoined twins with associated anomalies, especially cardiac abnormalities (mostly thoracopagus twins) carry poor prognosis.⁷ Complex fusion of the heart is considered ineligible for surgical separation and hence is not suitable for continuation of pregnancy. In some countries, termination of pregnancy can be offered to the family if severe forms of conjoined twins are diagnosed. It is clinically important to decide termination at an early gestational age as late-stage termination is fraught with problems.⁸ In Malaysia, termination of a pregnancy is illegal except when a registered medical practitioner does so if he or she “thinks, formed in good faith, that the continuation of the pregnancy would involve risk to the life of the pregnant woman, or injury to the mental or physical health of the pregnant woman, is greater than if the pregnancy was terminated”.⁹

In omphalopagus type of conjoined twins, the liver fusion occurs in 80% of the cases.¹ In this type of ventral union, the postoperative survival rate is high (82%) and with the presence of advances in surgery to separate the conjoined twins in recent years, this has been a factor for the obstetricians to suggest the continuation of the pregnancy.¹⁰ Thus, a discussion between the parents and the obstetrician had concluded that the pregnancy was going to be carried on since there was organ sharing of a lesser complexity. As a health professional in primary care, besides giving additional information on conjoined twins related complications (preterm birth, stillborn or early neonatal death, surgical delivery by caesarean section), the psychosocial issues need to be addressed during the antenatal visits. It is important to explore the mother’s feelings especially when knowing that her babies are not physically normal, tackle the parents’ anxiety, find out the burden that the parents might face and assess the ability to earn a living as they will need to spend most of the time commuting between home and hospital during and after the delivery. Parents and the medical team including all appropriate disciplines should negotiate the possible measures to be adopted to get a coordinated plan of care in this twin pregnancy. When the decision to continue with the pregnancy was made, many aspects needed to be worked on from the care of the mother throughout this pregnancy until the separation of the twins after delivery. Further discussion on the mode of delivery and surgical separation of the conjoined twins with the paediatric surgeon is crucial. In minimizing complications to the foetus and the

mother, the caesarean section is indicated as the choice of delivery.⁸ On the other hand, a successful separation requires adequate team management and meticulous preoperative planning.¹¹ Collaborative management involving multiple specialties play an important role in enhancing comprehensive antenatal care for these conjoined twins and preparing the best interest of the mother and her pregnancy.

It was not a surprise to get such a reaction from the parents after they were told about the conjoined twins. The worries of having a physically abnormal child were overwhelming at that time and they had the right to make such a decision. The doctor must respect the patient’s autonomy even though he carries a great responsibility in providing information about the benefits and risks of treatments related to this special pregnancy. Based on the facts given, the consultation aimed to avoid any decision that could have harmed the patient and give the best available option for the parents. The transparency of the discussion between the parents and medical health professionals is very important. These parents eventually agreed to continue the pregnancy after detailed explanations and discussion. In Malaysia, when there is a situation where the mother or parents insists on termination, medical considerations take precedence. No statute specifically related to conjoined twins exist, but there is some guidance in the Penal Code, Section 312.⁹ When the pregnancy poses a risk to the life of the pregnant mother or woman’s physical or mental health, a registered medical practitioner can make the decision based on his/her medical judgement. It has become a practice within the medical profession here that two doctors should agree before deciding to minimize the risk of any oversight.

Ethical approval about publication:

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