Two hearts, one rhythm: A case report on thoracoomphalopagus twins*

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ABSTRACT

A 21-year old woman, G1P0,was referred for further prenatal check-up with sonographic examination revealing conjoined twins at 29 weeks age of gestation. The fetuses were in breech presentation positioned face-to-face with fusion at the level of the thoraces and gastric bubble suggestive of thoracoomphalopagus twins. There was a definite communication between the two fetal circulations at the ventricular level as seen on fetal echocardiogram with a single cardiac rhythm shared between the two hearts. Close antenatal and fetal surveillance was done during the entire pregnancy duration. The patient was counseled about therapeutic options and explained of the complexity of their cardiac anatomy. The twins were delivered by cesarean section at 35 weeks due to preterm labor and a neonatal 2D-echocardiogram was done shortly after to re-assess their cardiac anatomy. Since the results revealed a shared ventricle, the twins were considered inseparable. The family was apprised of their poor prognosis and opted for natural death to occur.

Keywords: thoracoomphalopagus twins, conjoined twins

INTRODUCTION

Conjoined twins are a rare complication of monozygotic twins with a high incidence of morbidity and mortality.[1] Early identification and prenatal diagnosis allows adequate planning regarding possible management approaches, which can decrease maternal and fetal morbidity and mortality. Fetal prognosis however, greatly depends on the extent of fusion among the twins.

CASE

M.R., a 20 year-old primigravid, Filipino, single, unemployed, Roman Catholic from Talisay City, Cebu, admitted for the first time at a tertiary hospital for conjoined twins.

The patient's past medical history is unremarkable. Family history is significant for twinning on both sides. She has regular menstrual cycles lasting 3-5 days, using 2-3 pads per day. Coitarche was at 20 years old with 1 sexual partner. She is an occasional alcohol drinker and a non-smoker. She resigned from her job as a sales assistant at an optical shop when she learned she was carrying conjoined twins.

Antepartum prenatal care was started at 15 weeks age of gestation by last menstrual period at a Local Health Center in Argao, attended by a local midwife. However, at 19 weeks age of gestation, the patient's gravid abdomen was noted to be large for gestational age thus, an ultrasound scan was requested.

On ultrasonography at 22 weeks age of gestation, a multifetal intrauterine pregnancy with fusion at the thoracic and abdominal areas was noted suggestive of conjoined thoracoomphalopagus twins. She was then referred to this institution for further management.

The patient had her first prenatal checkup at our institution at 22-23 weeks age of gestation by ultrasound. Initial laboratories were requested and she was referred to a perinatologist. On examination, the abdomen was gravid with a fundic height of 30cm and an estimated combined fetal weight of 2.8-3.0 kg. Fetal heart tones were at 140bpm heard best at the left lower quadrant.

At 29 weeks age of gestation by first ultrasound, patient underwent a Congenital Anomaly Scan confirming the findings of conjoined thoracoomphalopagus twins. Their fetal heads were separate and facing each other with their fetal hearts appearing to be fused (Figure 1). They were in breech presentation and ventrally fused at the level of the thoraces and abdomen. They had separate upper and lower extremities, kidneys and male external genitalia and appeared to each have a gastric bubble and hepatic vessels with a fused liver (Figure 2). There was only 1 umbilical 4-vessel cord seen (Figure 3).

The scan further showed a complex configuration of the fetal hearts hence a fetal echocardiogram was done the following day. The results showed a definite communication between the two fetal circulations at the ventricular level. The right fetal heart (Twin A) shows a large atrial septal defect with a single AV valve and bridging leaflet draining into a single ventricle. The left fetal heart (Twin B) shows four cardiac chambers with a ventricular

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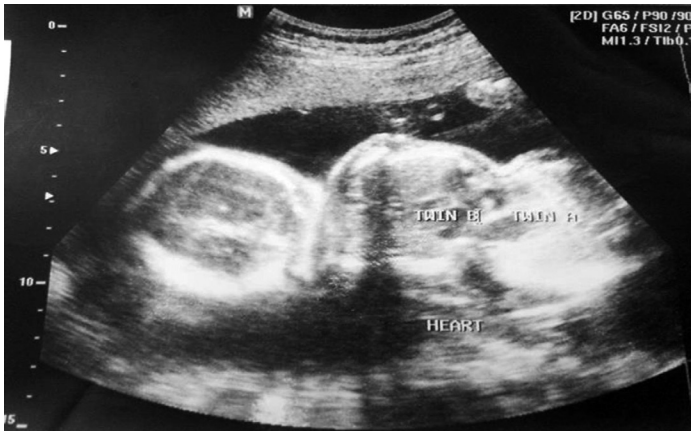

Figure 1. Ultrasound revealing separate fetal heads, which are facing each other and fetal hearts which appear to be fused

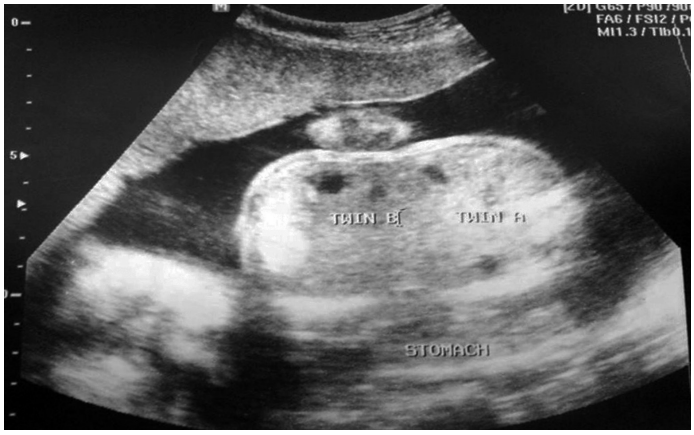

Figure 2. Separate gastric bubbles with a fused liver depicted on ultrasound

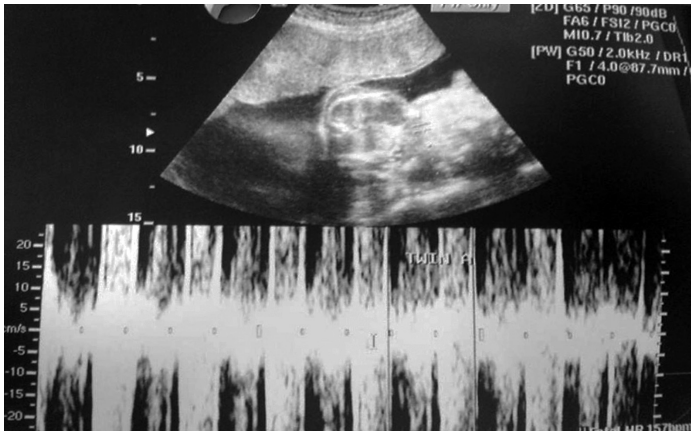

Figure 3. Ultrasonography showing only 1 cardiac pulsation for the twins and a single umbilical 4-vessel cord

septal defect with one of the great arteries providing a branch to the right fetal circulation (Figure 4).

The patient was informed of the possible extent of twin fusion and the chances of survival and separation. She has been having irregular premature uterine contractions for the past couple of days, thus she was admitted at our institution for steroid administration and monitoring for

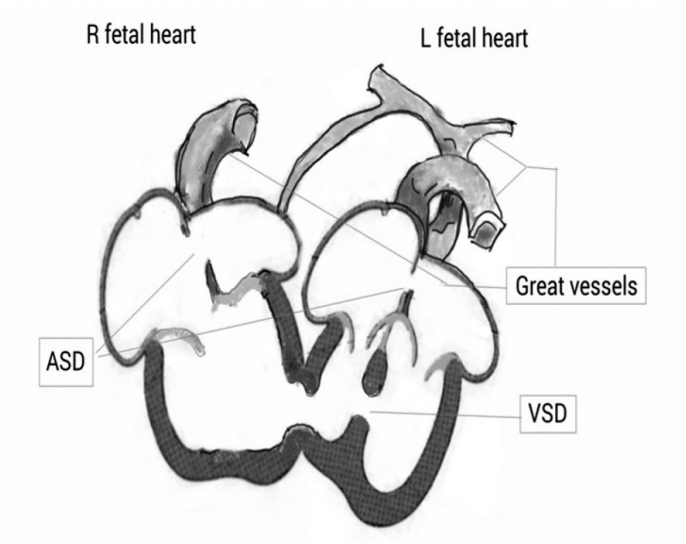

Figure 4. Antenatal Fetal Echocardiogram (March 4, 2015) showing a definite communication between the two fetal circulations at the ventricular level. The right fetal heart (Twin A) shows a large atrial septal defect with a single AV valve and bridging leaflet draining into a single ventricle. The left fetal heart (Twin B) shows four cardiac chambers with a ventricular septal defect with one of the great arteries providing a branch to the right fetal circulation.

the progress of labor. On admission however, there were no regular uterine contractions noted.

At 30 1/7 weeks age of gestation by first ultrasound, the patient and her partner met up with a pediatric geneticist for a closed door counseling. They were advised regarding the occurrence of conjoined twins, possible etiology and several things to expect as they go through with the pregnancy.

The patient continued to have regular weekly prenatal check-ups where progression of polyhydramnios was monitored as well as onset of uterine contractions, fetal growth and signs of fetal deterioration. Necessary blood products were being secured and coordination with other specialties such as pediatrics, anesthesia and surgery was also being facilitated.

A multidisciplinary meeting was held two weeks before the scheduled cesarean section attended by a pediatric cardiologist, pediatric anesthesiologist, pediatric intensivist, cardiologist-anesthesiologist, geneticist, a bioethics expert, obstetricians and pediatricians. A planned classical cesarean section was agreed upon at 37 weeks age of gestation. Issues on aggressiveness, feasibility as well as the bioethics of surgical separation were discussed. The pediatric cardiologist believes that because of the complexity of the cardiac fusion, chances of separation would be close to none. The consensus of the multidisciplinary team was that successful separation of the twins would be unlikely due to the complicated cardiac anatomy and that supportive care would be the

best approach at this point. The acute stage goal would be to stabilize the twins first once they were born and reassess the plan if they survive for more than 48 hours.

At 35-36 weeks age of gestation by ultrasound, the patient developed preterm labor. On physical examination, she was conscious, coherent with stable vital signs. Abdomen was gravid, with a fundic height of 36 cm and an estimated combined fetal weight of 4-4.2 kg. On Leopold's maneuver, Leopold's 1 was breech, Leopold's 2 cephalic, Leopold's 3 fetal backs on each maternal side and Leopold's 4 presenting part was unengaged. There were uterine contractions on abdominal palpation mild to moderate occurring every 3-5 minutes, lasting for 45-60 seconds. On internal examination, the cervix was dilated to 3 cm, 50% effaced, station -3 with intact membranes.

The admitting impression was G1P0 Pregnancy uterine 36 1/7 weeks age of gestation by early ultrasound, multifetal pregnancy, in preterm labor; Thoracoomphalopagus conjoined twins.

The patient was immediately scheduled for an emergency classical cesarean section and delivered live preterm male neonates with a combined weight of 4,300 grams, APGAR score of 9,10, Ballard's score of 36 weeks, appropriate for gestational age. Placenta was noted to be monochorionic, monoamnionic with its umbilical cord having four vessels identified to be 2 arteries and 2 veins. The twins were born facing each other and joined from the level of the nipples down to the level of the umbilicus. They have separate upper and lower extremities and genitalia. Estimated blood loss was 750 cc and one unit PRBC was transfused postoperatively.

The mother's course in the ward was unremarkable and she was discharged on the 4th hospital day.

For the twins on the other hand, after their delivery, IV fluids were started, initial newborn care was provided and they were kept thermoregulated. Because of tachypnea, O2 inhalation via nasal prong at 3LPM was given. OGT feeding was started at 10 cc every 3 hours with increments of 2cc after each 2 successful feedings. A neonatal 2D echo was done revealing a single common atrium with slight narrowing in between. The posterior ventricle of Twin 1 gives off the aorta with its aortic arch coursing to the right while the posterior ventricle of Twin 2 appears to have a ventricular septal defect as its outlet. A middle ventricle shared by both twins were noted which gives off the aorta of Twin 2 and both pulmonary arteries of Twin 1 and 2. It also communicates with the posterior ventricle of Twin 1 through a ventricular septal defect (Figure 5).

The family was apprised of their poor prognosis due to the ventricular fusion and a "do not resuscitate" waiver was signed. After 4 days, the twins succumbed and died with the final diagnosis of: (1) Preterm Male Neonate delivered via classical cesarean section secondary to Conjoined Twins BS 36 weeks AOG, BW 4,300 grams, AGA; (2) Complex Congenital Heart Disease.

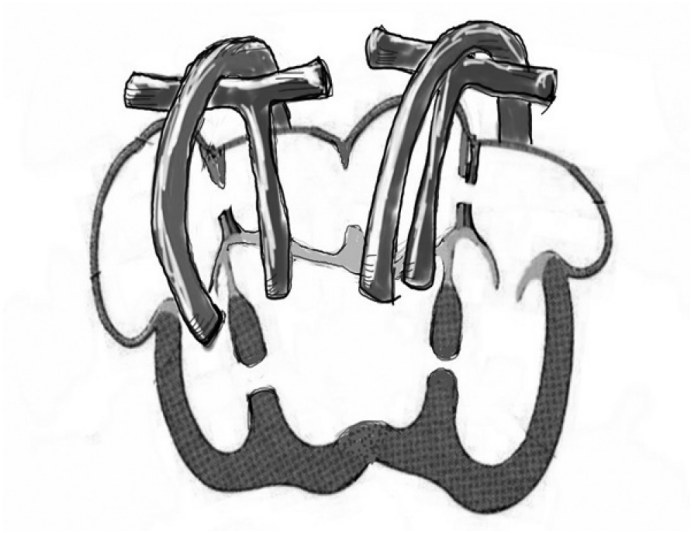

Figure 5. Neonatal 2D-Echocardiogram (April 25, 2015) showing a single common atrium with slight narrowing in between. The posterior ventricle of Twin I gives off the aorta with its aortic arch coursing to the right while the posterior ventricle of Twin 2 appears to have a ventricular septal defect as its outlet. A middle ventricle shared by both twins were noted which gives off the aorta of Twin 2 and both pulmonary arteries of Twin 1 and 2. It also communicates with the posterior ventricle of Twin 1 through a ventricular septal defect.

DISCUSSION

Conjoined twins represent one of the rare forms of twin gestation with incidence of 1 in 50,000 live births[2]. They are classified according to the site of fusion with thoracoomphalopagus occurring most frequently (28%)[3]. This type of twinning involves union of two bodies fused from the upper chest to the lower chest. They usually share a heart and may also share the liver or part of the digestive system[4].

The etiology of conjoined twins is not known. In general, there are no karyotypic abnormalities, nor do race, heredity, birth order, or consanguinity appear to influence the process[5]. The malformation is believed to occur between the 13th-15th day from fertilization andtwo theories have been postulated explaining their etiology. The older theory called the fission theory, states that conjoined twins occur when a fertilized ovum begins to split into identical twins but is somehow interrupted during the process and develops into two partially formed individuals who are stuck together. The fusion theory on the other hand, believes that twins become conjoined after the fertilized ovum separates but stem cells, which seek out cells of the same type, find like cells on the other twin and fuse the twins together. When two newly-separated

identical twin embryos are lying in close proximity to one another, sometimes signals get mixed up and cells will attach to other cells of the same type that belong to the other twin[6].

Several cases of conjoined twins have been documented in the Philippines. Carl and Clarence Aguirre were born conjoined at the top of the head (vertical craniopagus) and a revolutionary new process was used in separating them as their doctors chose to separate them in smaller operations allowing them to recuperate after each surgery to decrease the rate of mortality and morbidity from marathon surgeries[7]. They were successfully separated in Montefiore Medical Center in New York after four major surgeries.

In April of 2003, a set of 9-month old female thoracoomphalopagus twins from the Philippines was successfully separated in a hospital in Taiwan. They were delivered by cesarean section after 38 weeks of gestation and were externally fused from the lower chest to the upper abdomen in the ventral aspect. After a full preoperative assessment including cardiac and abdominal echograms, plain film of the body, whole body computed tomography, chest to abdomen MRI, and upper gastrointestinal serial films, the twins received tissue expander implantation to allow ease in reconstruction[8]. One month after expansion surgery, a successful surgical separation was made.

Overall survival of conjoined twins is roughly around 25%[9]. Approximately half are stillborn and about 34% die within the first day of life. Survival depends upon the type of fusion and the presence of associated anomalies. Although there may be cardiac abnormalities in any type of fusion, thoracopagus twins have the highest incidence of abnormal cardiovascular findings with a 90% incidence of shared pericardium[10] and major myocardial connections in some 75% of cases[11]. In thoracoomphalopagus, the degree of fusion of the heart and intracardiac anatomy determines the prognosis. When a common heart is present, the chances for a successful surgical separation become negligible.

Cardiac anomalies in twins are classified by Leachman into Group A: separate hearts, separate pericardium; Group B: separate hearts, common pericardium; Group C: fused atria, separate ventricles; and Group D: atrial and ventricular fusion[12]. In the index case, a definite communication between the two fetal circulations at the ventricular level was seen with only 1 cardiac pulsation for both twins on congenital scan and fetal 2D echo, indicating cardiac fusion, which classifies them under group C. On neonatal 2D echo, however, there was atrial and ventricular fusion, which classifies them under group D. If only pericardium is shared between the hearts, they have electrographic complexes that occur at different times and rates hence could be readily separable whereas if they share atrial or ventricular tissue, their hearts depolarize simultaneously and the QRS complexes and arterial pulsations coincide.

The definite communication between the two fetal circulations at the ventricular level in the index case greatly affects treatment options in terms of separation because of this complex cardiac configuration. Hence, echocardiography is necessary as it allows accurate delineation of cardiac fusion, intracardiac anatomy and ventricular function. With the advent of more sophisticated imaging techniques such as MRI and Computed Tomography, echocardiography remains to be the bedrock of cardiac assessment as it provides an accurate, safe, non-invasive and easily portable method for cardiovascular system assessment[13].

Aside from the cardiac fusion seen in the index case, the fetuses also show fusion at the level of the gastric bubble and possible union of the liver. Hence, postnatal imaging studies including plain film, CT, MRI, and contrast GI studies are necessary to evaluate their condition and accurately define anatomic fusion, vascular anomalies and associated abnormalities, which are essential in planning and prognostic estimation[8]. Because of the complex cardiac anatomy, fusion of the stomach and possibly the liver, prognosis is generally poor for these twins.

Conjoined twins can be diagnosed using standard ultrasound as early as the first trimester. However, false-positive results are common before 10 weeks because early in gestation, monoamniotic twins may appear conjoined and fetal movements are limited[14].

A high index of suspicion for thoracoomphalopagus twins should be made in all monochorionic monoamnionic pregnancies if the twins maintain a constant and often unusual relative position more so, if the head and neck are constantly hyperextended[15]. Ultrasonographic identification of any of the following classical signs may suggest the diagnosis if both fetal heads are in the same plane, if there is unusual backward flexion of the cervical spine, no change in the relative position after maternal movement and manual manipulations, and inability to separate fetal bodies after careful observation[16]. Suspicion is also increased if maternal hydramnios and fetal malpresentation are present[17].

In the index case, the patient was diagnosed after 20 weeks gestation and presented with a gravid abdomen larger than expected for her gestational age. On ultrasonography, both fetal heads were in the same plane, with lordosis of both twins, and inability to separate the bodies after probe manipulation.

Very early prenatal diagnosis and first-trimester 3D imaging provide little additional practical medical information compared to the 11-14 weeks' ultrasound

examination[14]. Thus, a definite diagnosis should be made with caution, after meticulous sonographic examination at this early stage[18].

If an ultrasound detects conjoined twins, a more detailed ultrasound and an echocardiogram should be used about halfway through pregnancy to better determine the extent of the twins' connection and organ functioning[14]. In the index case, initial ultrasound was done during the 3rd trimester of pregnancy detecting thoracoomphalopagus twins. A congenital anomaly scan and fetal echocardiogram were done thereafter to better delineate the extent of organ fusion reducing the chances for a false positive result.

Preterm labor is also common among conjoined twins due to the rapidly increasing abdomen from increasing fetal size and polyhydramnios. This increase in amniotic fluid volume, which was also seen in the index case, may be indicative of other underlying congenital anomalies along the GI tract or a possible twin-to-twin transfusion syndrome. However, the latter may be ruled out as no fetal discordancy was noted on ultrasound. For cases of polyhydramnios, amnioreduction may be done to control preterm labor and alleviate dyspnea. In the index case, the patient was given steroids to aide in fetal lung maturity when she developed preterm labor to allow for further diagnostic tests as the extent of cardiac fusion was yet to be determined as of this point.

After diagnosis, timing and mode of delivery should be planned based on the possibility of survival, size, nature of the fusion and parental wishes. Scheduled delivery in a tertiary care center is ideal so that procedures required to evaluate the twins can be carried out shortly after birth[19]. An elective cesarean section at term ensures the best chance of survival.

Cesarean delivery is recommended in most third-trimester deliveries because of the high incidence of dystocia and resultant fetal damage. Vaginal delivery should be reserved for early stillbirths and for conjoined twins that are incompatible with life[19] provided that there is no cephalopelvic disproportion. Elective cesarean delivery should be performed near term after confirmation of fetal lung maturity. Twin delivery may otherwise lead to overdistension, uterine atony[20] and possibly, uterine rupture secondary to overdistension of the uterus. In the case presented, planned mode of delivery was by cesarean section with a classical incision to prevent maternal and fetal trauma. Blood products were prepared in case of hemorrhagic complications brought about by uterine overdistension. The patient was also apprised of possible future pregnancies and the need for spacing, as her succeeding pregnancies will be delivered by cesarean section due to the high likelihood of uterine rupture with this type of incision.

As most parents opt for immediate termination of pregnancy at confirmation of the diagnosis, there has been limited data on the prenatal follow-up of conjoined twins. And when parents opt for conservative management on the other hand, half of the fetuses usually die in utero and another 44% die during the neonatal period. Generally however, the main goal of prenatal management of conjoined fetuses includes maximizing the potential for their survival and minimizing maternal morbidity[21].

Pediatric involvement is of equal importance in the management of conjoined twins as they will be receiving and managing a potentially morbid set of twins. After birth, evaluation of both twins will be conducted to assess the extent of organ system sharing. Referral to a pediatric surgeon early in the course of her treatment can help in the preoperative evaluation of the case. Timing of separation and separation plan should be individualized according to the need of emergent separation and the degree of organ fusion.

Majority of case reports on conjoined thoracoomphalopagus twins were identified early in pregnancy (<12 weeks) with parents usually opting for termination. However, with the case presented, presence of conjoined twins were noted at 21-22 weeks AOG and termination of pregnancy is not an option as it is illegal in our country. The Philippine Society of Obstetrics and Gynecology (POGS) advocates respect for human life starting from conception and consider it unethical to terminate a pregnancy because of fetal defects that have been detected on ultrasound or invasive fetal testing[22]. Other ethical issues which pose a dilemma in planning out the treatment approach and management of conjoined twins would be whether to administer steroids for fetal lung maturity in a set of twins which have a poor prognosis and the likelihood of having to choose between saving one over the other.

The principle of double effect must be considered in handling cases of conjoined twins as this provides us with a set of guidelines for determining when it is morally permissible to perform an action in pursuit of a good end in full knowledge that the action will also bring about bad results. Four conditions should be met if the action in question is to be ethically permissible: (1) that the action contemplated be in itself morally good or morally indifferent, (2) that the bad result not be directly intended, (3) that the good results not be a direct causal result of the bad result, and (4) that the good result be proportionate to the bad results[23].

In the case presented above, the family was apprised of the guarded condition of the twins and their poor prognosis. Their decision to allow natural death to occur was respected.

SUMMARY

Options for the treatment of thoracoomphalopagus twins are largely dependent on the anatomy of the cardiovascular system while separation of the liver and GITs can be managed readily in most cases.

Several successful surgical separations for thoracoomphalopagus twins have been documented however none involved ventricular fusion. Among those with ventricular involvement with attempted surgical separation, none survived. Therefore, the case presented above has a very poor prognosis in terms of survival and successful separation. Safe delivery for the mother and fetuses as well as supportive care for the twins remained to be the main objective during the entire course. ■

REFERENCES

1. Shepherd, L.J., et. al Conjoined Twins in a Triplet Pregnancy: A case report. Obstetrics and Gynecology, September 6, 2011.
2. Osmanagaoglu MA, TurhanAran, SuleymanGuven, Cavit Kart, OzgurOzdemir, HasanBozkaya. Thoracopagus Conjoined Twins: A Case Report. *ISRN Obstet Gynecol* 2011; 2011:238360.
3. Kaufman MH.The embryology of conjoined twins. *Childs Nerv Syst* 2004 Aug; 20(8-9):508-25.
4. http://en.wikipedia.org/wiki/Conjoined_twins
5. Cuillier F, Dilion KC, Grochal F, Scemama JM, Gervais T, Cerekja A, Piazze J. Conjoined twins: what ultrasound may add to management. *J Prenat Med* 2012 Jan-Mar; 6(1):4-6.
6. Le Tao, VikasBhushan, Neil Vasan (2010). First Aid for the USMLE Step 1: 2010 20th Anniversary Edition. USA: The McGraw-Hill Companies, Inc. ISBN 978-0-07-163340-6.
7. Fierro PP. Conjoined Twins Profile: Carl and Clarence Aguirre. About Health, 2015.
8. Cheng-Hui Chiu, Andy Shau-Bin Chou, Chau-Chin Lee, Sea-Kita Lee, Pau-Nyen Chong. Imaging in the Preoperative Assessment of Omphalopagus Conjoined Twins-A Case Report. *Tzu Chi Med J* 2006; 18:61-64.
9. Stone JL, Goodrich JT. The craniopagus malformation: classification and implications for surgical separation. DOI: 1084-1095: Apr 2006.
10. Rudolph AJ, Michaels JO, Nichols BL. Obstetric management of conjoined twins. Birth defects original article series, 1967.
11. Leachman RD. Cardiovascular evaluation of conjoined twins. Birth defects original article series, 1967.
12. Leachman RD, Latson JR, Kohler CM, McNamara DG. Cardiovascular evaluation of conjoined twins. Birth Defects 1967; 3:52-65.
13. Andrews RE, McMahon CJ, Yates SC, de Leval MR, Kiely EM, Spitz L, Sullivan ID. Congenital heart disease: Echocardiographic assessment of conjoined twins. Heart 2006; 92:382-387.
14. Pajkrt E, Jauniaux E. First Trimester Diagnosis of Conjoined Twins. Prenatal Diagnosis. September 2005; vol 25, issue 9.
15. Sharma, UK, et al. Antenatal Detection of Conjoined Twins, Case Report. J. Nepal Assoc, 2007.
16. Kalchbrenner M, Weiner S, Templeton J, Losure TA. Prenatal Ultrasound Diagnosis of Thoracphagus Conjoined Twins. *J Clin. Ultrasound* 1987; vol 15, issue 1.
17. Nelson MM, Bhettay E, Beighton P. Excessive Siamese Twinning in South Africa. S. Afr. Med. J., 50, 697 (April, 1976).
18. Bornstein E, Santos R, Timor-Tritsch IE, Monteagudo A. "Brothers in Arms": 3-Dimensional Sonographic Findings in a First-Trimester Thoraco-Omphalopagus Conjoined Twin Pair. JUM January 1, 2009 vol. 28 no. 1 97-99.
19. Norris C, Finberg HJ, Peoples W, Nielsen ML, Tiller GR. Twins, Conjoined, Thoracoomphalopagus. The Fetus.net 1994-08-12-11.
20. Khalid Kamal, Abdulhamid Ibrahim, Roxas Renato, ShahbazSarwar CM, Al Rabeeah A. Conjoined Twins Treatment & Management, Medscape, 2012.
21. ApostolosAthanasiadis, et al. Prenatal Diagnosis and Management Conditions of Conjoined Twins. Donald School Journal of Ultrasound in Obstetrics and Gynecology, Jan-March 2007.
22. Philippine Obsterical and Gynecological Society (Foundation) Inc. (POGS) Committee on Ethics and Medical Practice. Ethical Guidelines in Obstetrical and Gynecological Practice, Medical Education and Research 2004: Manila.
23. Jacobs, Richard M.The Principle of Double Effect. MPA 8300 Leadership Ethics, Villanova University; July 7, 2013.