Case Report

A Case of Dicephalus Parapagus Dibrachius Dipedis Twins with Review of Literature

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Abstract

Conjoined twins are embryologic abnormalities which occur due to incomplete division of monozygotic twins. They are classified based on the most prominent site of union. Our aim was to use a postmortem examination to allow for proper classification and description of various coexisting pathologies present within the conjoined twins. We report an occurrence of a dicephalus parapagus dibrachius dipedis conjoined twin delivered to a 28-year-old female at our hospital. An autopsy was performed on the twin following an early demise. The detailed morphological examination findings with emphasis on the unique cardiovascular anatomical finding are reported along with a review of relevant literature. This report illustrates the occurrence of this variant of conjoined twins and the need to establish detailed institutional/national registries for birth defects and congenital anomalies in developing countries.

Keywords: Anomalies, autopsy, conjoined twins, parapagus

Received on: 09-06-19 **Review completed on:** 27-06-19 **Accepted on:** 12-09-19 **Published on:** 06-12-19

Introduction

Conjoined twins are embryologic abnormalities which occur due to incomplete division of monozygotic twins. Their etiology is uncertain. The occurrence of conjoined twins is not peculiar to humans as it has been documented in fish, reptiles, birds, and other primates.[1]

The most famous conjoined twins were "Eng and Chung Bunker" (1811–1874) born in the Kingdom of Siam (now Thailand).[2] The often-used phrase "Siamese twins" for conjoint twins was coined as a reference to "Eng and Chang."

There is a variable prevalence of conjoined twins worldwide. It has been estimated to be 1 in 50,000 pregnancies but 1 in 200,000 live births.[3] Reports have, however, cited the prevalence as high as 1 in 2,800 live births in India[4] and as low as 1 in 200,000 live births in the United States.[5] In the largest study on conjoined twins carried out, Mutchinick *et al*. reported the total prevalence of conjoined twins as 1.47/100,000 births worldwide.[6] There is a female predominance (ratio: 2:1) among conjoined twins.[6]

In Nigeria, West Africa, the incidence of conjoined twins is not known. The earliest report was in Sokoto, Northern Nigeria, 1935, of a 25-year-old para 3 woman who had a home delivery.[7] The conjoined twins reportedly shared only abdomen and skin but no internal organs.[7] They were separated by a British missionary doctor.[7]

Since the first conjoined twins in Nigeria were reported in 1936,[7] nineteen cases have so far been documented in the Nigerian medical literature.[8] Most of the cases have come from Zaria in Northern Nigeria (28%).[9] Other cases have been reported from Ife[10] and Enugu.[11] It may be pertinent to note that there are cases of conjoined twin reports that have never made the medical literature.

At the University College Hospital, Ibadan, two previous cases of conjoined twins have been reported.[12] The first case was by Gupta and involved a set of pyopagus conjoined twins,[13] and the second case as reported by Omokhodion *et al*. involved a pair of thoracopagus conjoined twins.[12]

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Access this article online

Quick Response Code:

Website: www.atpjournal.org

DOI: 10.4103/atp.atp_20_19

How to cite this article: Nweke MC, Adesina OA, Akinola A, Iyapo O, Ogun GO. A case of dicephalus parapagus dibrachius dipedis twins with review of literature. Ann Trop Pathol 2019;10:166-71.

In this article, we present the autopsy findings of a conjoint twin emphasizing the arrangement of viscera in the thoracic and abdominal cavities along with the clinical presentation and review of literature.

Case History

A 28-year-old gravida 3 para 2+0 (with one living child) and two previous cesarean sections presented at the antenatal clinic of the University College Hospital, Ibadan, Nigeria, at the gestational age of 35 weeks with an ultrasound report of a conjoined twin pregnancy. She started antenatal care at a private hospital at a gestational age of 20 weeks but did not do an anomaly ultrasound scan earlier.

The first ultrasound report of conjoined twins was at a gestational age of 28 weeks (it revealed a conjoined twin with two heads, two upper limbs, and two lower limbs). This necessitated a second obstetric ultrasound opinion which confirmed a dicephalic parapagus twin. She did not represent till gestational age of 36 weeks. There was no history of ingestion of teratogens or alcohol during the pregnancy. There was a family history of twinning but no history of a previous child with a congenital anomaly.

She had an emergency classical cesarean section on account of premature labor. The intraoperative findings were: a live conjoined female twin delivered and weighing 3.9 kg with Apgar scores of 2 in the 1st min and 4 in the 5th min. There was monochorionic, monoamniotic placenta which was normal on gross examination. The conjoined twins suffered an early neonatal death after 30 min of active resuscitation.

Postmortem findings

An external examination is that of a set of conjoined twins having two heads attached at the neck on a single torso bearing a pair of well-developed upper and lower limbs and a single umbilical opening with a female introitus. Both heads show marked central and peripheral cyanosis. The conjoined twins weigh 3.910 kg. The anthropometric measurements of the conjoined twins are described in Table 1.

Figure 1 shows the fused ribcage of the joint twins.

Figure 2 illustrates the major paired internal organs with petechial hemorrhages on the surface of the organs.

Cardiovascular system examination showed two maldeveloped partly fused hearts. Both hearts have a common primitive atrium which opens into a single-chambered ventricle in Twin A and a double-chambered ventricle in Twin B. This common atrium has neither left-sided nor right-sided morphology. The single-chambered ventricle in Twin A has no accompanying hypoplastic ventricle and is solitary. The atrioventricular valve shows continuity with the pulmonary valve. There is a pulmonary artery that arises from the ventricle of Twin A that opens into the pulmonary vasculature. There is an intramuscular ventricular septal defect involving the two-chambered ventricle of Twin B allowing communication of the left and right ventricles of Twin B. The ventricular septal defect measures 1.4 cm. There is a single aorta which arises from the left ventricle of the heart of Twin B that descends to form the abdominal aorta from which paired vessels supplying the intra-abdominal viscera arise. The coronary arteries supplying Twin B (left heart) arise from this aorta. There is also a pulmonary artery which arises from the right ventricle of Twin B which goes on to supply the lungs of Twin B through the pulmonary vasculature. There was no ductus arteriosus observed.

Figure 3 illustrates cardiovascular defects.

There are two separate well-developed airways. The mucosa of each of the airways (trachea and larynx) shows marked congestion and petechial hemorrhage from intubation and resuscitation. Both tracheas bifurcate into the left and right bronchi that also open into two pairs of hypoplastic lungs. The pleural surfaces show petechial hemorrhage. Cut surfaces show mild congestion. The floatation test carried out in each of the lung zones of both lungs of Twin A and Twin B is negative as biopsies from all segments of the lung floated, indicating some inflation of both lungs at birth.

There are two separate livers each with a smooth Glisson capsule. The smaller liver is retroperitoneal in location. The larger liver is well developed with a distinct hepatobiliary system, whereas the smaller liver shows an incompletely developed hepatobiliary system with no gallbladder identified. This is seen histologically as primitive hepatocytes with paucity of biliary ducts/canaliculi. The distinct hepatobiliary systems of both livers open through a single duct at the ampulla of Vater

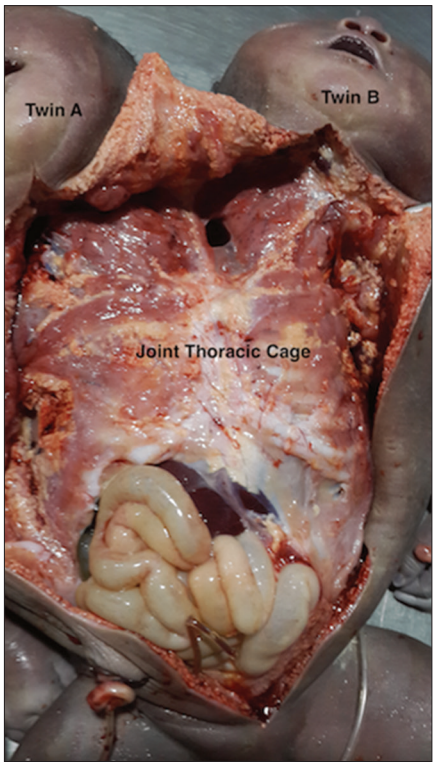

Figure 1: Joint thoracic cage of conjoined twins

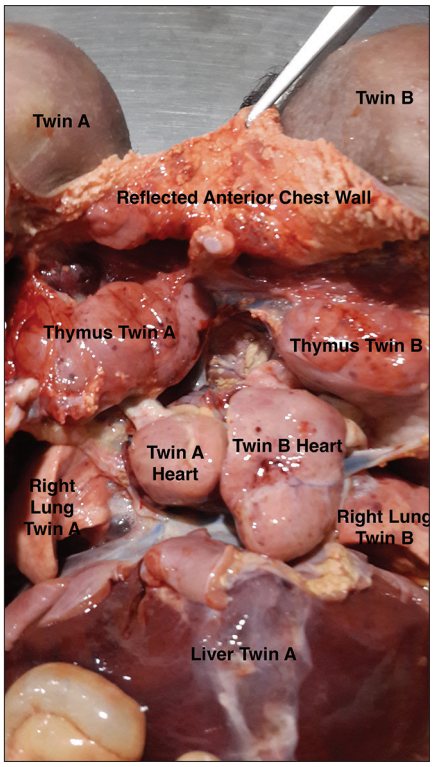

Figure 2: *In situ* arrangement of the various organs of the conjoined twins

into the second part of the duodenum. The gallbladder of the larger liver contains bile, and the hepatobiliary tract is patent.

There are two sets of well-developed thyroid tissue seen. Cut sections are normal. There are two sets of well-developed pancreatic tissue, which unite distally and open through a common duct through the ampulla of Vater in the second part of the duodenum.

Two separate well-developed thymic organs are seen, and their cut sections show a normal pale-brown appearance.

Two well-formed sets of esophagi are seen which open separately into separate well-formed stomach sacs. There is no demonstrable tracheoesophageal fistula/anomaly. Each of the stomach sacs opens into a common duodenum, which continues into the jejunum. The gastrointestinal tract then continues into the ileum, ascending colon, and transverse colon, which appear grossly unremarkable. The descending colon/rectum shows gangrenous bowel loops. The anus is patent with meconium seen within the canal.

The scalp, skull, and dura of each twin appear grossly intact.

There is extensive subgaleal petechial and purpuric hemorrhage involving the scalp and the skull of both twins. The brains of the individual twins are within normal limits and show prominent subarachnoid hemorrhage. Cut sections through both cerebral lobes show intracerebral hemorrhage with intraventricular extension.

Single organs common to the conjoint twins include a solitary spleen and a pair of well-developed right and left kidneys.

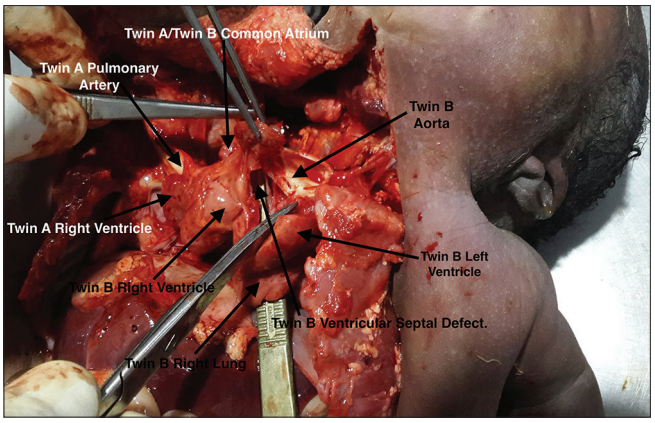

Figure 3: Detailed illustration of cardiovascular pathology

Table 1: All postmortem anthropometric measurements/organ weights in each of the individual conjoint twins

Combined weight (kg)	3.91
Occipitofrontal circumference (Twin A) (cm)	36.5
Occipitofrontal circumference (Twin B) (cm)	34
Combined neck circumference (cm)	39
Chest circumference (cm)	40
Crown-rump length (Twin A) (cm)	37
Crown-heel length (Twin A) (cm)	53
Crown-rump length (Twin B) (cm)	34
Crown-heel length (Twin B) (cm)	53
Combined heart weight (g)	50
Ventricular septal defect (cm)	1.4
Both sets of lungs (Twin A/Twin B) (g)	25
Larger liver/smaller liver (g)	40/15
Thyroid (g)	10
Thymus (Twin A)/thymus (Twin B) (g)	30/35
Spleen (g)	25/25
Both kidneys (g)	25/25
Brain (Twin A)/brain (Twin B) (g)	40/40

These organs appear normal. The capsules of both kidneys strip with ease to reveal a smooth subcapsular surface with prominent fetal lobulation. Cut sections show good corticomedullary differentiation. Both kidneys open through single well-formed ureters into the apical surface of the urinary bladder. The ureters appear grossly unremarkable. The urachus is still identifiable. The mucosal surface of the bladder shows mild congestion.

In the musculoskeletal system, there were two separate vertebral columns which are fused distally at the lumbar region [Figure 4] with an intervening intervertebral concavity which manifests as a gibbus dorsally.

Discussion

Conjoined twins embryologically arise from an aberration of monozygotic monoamniotic twinning due to incomplete late division (postday 13) of a monozygotic embryo at Carnegie

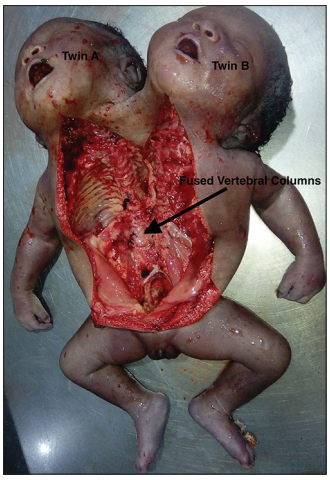

Figure 4: Fused vertebral columns of the conjoined twins

Stage 6 (days 12–15) after formation of the embryonic disc and rudimentary amniotic sac. At this stage, the complete separation of the inner cell mass within the chorionic mass does not occur. As a result, the nonseparated parts of the otherwise normal twins remain fused throughout development.

Two opposing theories have been proposed to elucidate the sequence of events in the pathogenesis of conjoined twins.[6] They are named as the "fission" and "fusion" theories.[6] The fission theory explains that conjoined twins arise from the fission of a zygote very early in their embryonic development. It arises from a simple molecular disorder at a deep cellular level distorting cellular adhesion or apoptosis in a very early stage of embryogenesis, thus resulting in an incomplete split of the inner cell mass.[14] The fusion theory hypothesizes that conjoined twins arise from fusion of stem cells of monozygotic embryos early in their development.

Proponents of the fusion theory propose that with the exception of parapagus conjoined twins, all conjoined twins can be attributed to the fusion of two separated embryos.[15] They argue "that no theoretical fission of the vertebrate embryo at any stage of the development, in any plane, in any direction can explain the selection of the observed sites of fusion, details of the union or the limitation to specific areas in which the twins are found to be joined." Peter *et al*. in support of the fission theory explain that the incidence of mirror imaging should be the same in all monoamniotic twins whether conjoined or not, if fusion accounted for all cases of conjoined twins.[16] The observation that the incidence of mirror imaging is higher in conjoined twins than in separate twins proves that the fusion theory cannot be correct.[16]

Conjoined twins are classified based on the most prominent site of union which is accompanied by the suffix-pagus (meaning fixed). The various types include cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pyopagus, rachipagus, and asymmetric conjoined twins.[6] Cephalopagus conjoined twins have two faces and are joined at the tip of the head.[6] Thoracopagus conjoined twins are joined face to face from the upper thorax to the upper part of the abdomen, always involving the heart.[6] Parapagus twins are laterally joined and regularly share the pelvis.[6] There are different variations of parapagus twins, i.e., dithoracic parapagus (separated thoraces), parapagus dicephalus (one trunk and two separate heads), and parapagus diprosopus (one trunk, one head, and two faces) to name a few.[6] Omphalopagus conjoined twins involve fusion at the umbilicus region frequently at the site of the lower thorax but never the heart.[6] In ischiopagus conjoined twins, the union includes the lower abdomen with duplicated fused pelvic bones, and the external genitalia are always involved.[6] Pyopagus is dorsally fused over the perineal and sacrococcygeal areas.[6] They have two rectums but one anus.[6] Rachipagus involves fusion of the dorso-lumbar vertebral columns and rarely the cervical vertebrae and occipital bone.[6] Craniopagus conjoined twins also referred to as helmet conjoined twins are joined by the skull and share meninges but rarely share the brain surface.[6] They do not share attachment over the face and trunk.[6]

Asymmetric conjoined twinning is a term used by some authors to refer to parasitic conjoined twins and fetus in a fetus.[6] Parasitic twins occur when one embryo of a pair of monozygotic twins starts to develop, with failure to fully separate.[6] As a result, one embryo's development prevails over the other. Some authors do not consider them as a type of conjoined twins as the embryo involved is incompletely formed and wholly dependent on the body functions of the complete fetus.[6] Parasitic twins are reported to have a prevalence rate twenty times less frequent corresponding prevalence rates in symmetrical types of conjoined twins.[17]

These various types of conjoined twins may be divided into two dorsal and ventral subgroups based on the aspect of the embryonic disc involved. Ventral subgroups are united over a single yolk sac along with a shared abdomen and umbilicus. Ventral unions are more common (87%) and can occur rostrally (48%), laterally (28%), and caudally (11%).[18] The dorsal subgroups are rare (13%), conjoined in the neural tube, and have a separate abdomen and umbilical cord.[18] This subgrouping may have an impact on the prognosis, as it determines the severity of visceral malformation.[19]

Among all the variants, the thoracopagus variety accounts for the most common (42%) of reported cases.[6] The parapagus dicephalus variant of conjoined twins is the second most common at 11.5%.[6] This is followed by craniopagus and omphalopagus at 5.5% each.[6] Other types of conjoined twins such as ischiopagus, rachipagus, parapagus diprosopus, and pyopagus were observed in <3% of cases.[6] Thoracopagus type is almost four times more frequent in females than in males.[6]

This case report involves dicephalic parapagus dibrachius dipedis conjoined twins with peculiar cardiovascular pathology. This is important as the heart is a critical organ for survival if separation of the conjoint twins is to be considered

for postnatal survival. Both hearts showed a common primitive atrium which opened into a single-chambered ventricle in Twin A and a double-chambered ventricle in Twin B. Previous studies have described fused hearts with complex anatomy.[20,21]

In her review, Enid Gilbert Barness opined that cardiac morphogenesis in conjoined twins is dependent on the side of the conjoined fusion along with the temporal and spatial influences.[20] This determines morphogenesis along with abnormally oriented embryonic axes.[20]

Seo *et al*. introduced a system of classification of cardiovascular system anomalies among conjoined twins using the degree of fusion and symmetry of hearts and great vessels.[22] This classification is divided into five types.[22]

- Type 1 involves cases with no vascular union on cardiac, aortic, and inferior vena cava levels
- Type II includes cases with separate vessels with union between aortas and inferior vena cava
- Cases with cardiac fusion at the atrial level are classified as type III. This Type III group can further be divided into two groups: Type IIIa in which there is a right atrial fusion and Type IIIb where there is a theoretical fusion between the left atria or between the left and right atria
- Type IV represents the fusion of both atria and ventricles irrespective of the number of chambers
- Type V represents a single heart in one of the twins.

Using this classification, our index case can be categorized into Type IV. Jeong Wook Seo's classification also states that the external morphologic type and the situs of each twin pair were closely related to the cardiac abnormalities.[22] This has also been proven to be correct in other studies.

Dicephalus and thoracopagus conjoint twins were more likely to be associated with abnormal situs and complex cardiac fusion.[22] This results from disturbed cross-signaling between tissues in adjacent primitive streaks, especially at the rostral aspect of the early embryonic disc. The right twin heart is usually more severely malformed with possible defects of laterality or situs inversus of the right twin organs often accompanying the heart abnormalities.[21,23]

The two hearts present suggest early separation of the anterior ends of the early embryonic axes as the heart develops from cardiogenic mesoderm cranial to the notochord. The fused common atrium suggests juxtaposition of the hearts at the time of formation of the atria. However, this occurs before the definitive pleural and pericardial cavities are formed.

Our index case had a female sexual phenotype, which is in keeping with the predominant female occurrence among conjoint twins.[20] However, the occurrence of the parapagus type is more common in males.[6] It is worthy to note that despite the external phenotype seen in our index case, no female reproductive organs were identified even after extensive histological sampling for confirmation. Neural tube defects and genitourinary anomalies have been observed in >15% of dicephalus and diprosopus types.[6] There have been previous reports worldwide of dicephalic conjoined twins presenting as either tetrabrachius or dibrachius.[24] However, none has been documented in Nigerian medical literature to the authors' knowledge.

There are no records present in the literature of a familial history of conjoined twins or its associations with other unrelated anomalies.[6] There has also been no conclusive evidence documented regarding the causative effects of radiation, medication, or environmental agents.[6] The mother in our case did not have any family history of conjoined twins or any history of previous births with congenital anomaly. There was also no history of the use of known teratogenic agents during the course of her pregnancy. A first-trimester ultrasound screening to rule out congenital anomaly is important in the early diagnosis of conjoined twins. High-resolution vaginal sonography makes diagnosis possible as early as the 8th week.[25] Thus, in good hands, ultrasound prenatal diagnosis is relatively easy and reliable.[25] This was, however, not carried out in the mother in contrast to the usual dictates of routine antenatal care.

Prognosis is very poor among conjoined twins. They are generally incompatible with life; 65% of cases are stillborn, while of those born alive, 35% die within the first 24 h. The prognosis is highly dependent on the type of fusion with associated structural defects.[19] In our case, death occurred roughly 20 min after delivery.

The method of delivery would depend on prenatal assessment. Cesarean section is recommended in most third-trimester deliveries, whereas vaginal deliveries are reserved for stillbirths and forms of conjoined twins incompatible with life.[26] In our index case, the late presentation by our patient and the preterm onset of labor necessitated an emergency cesarean section.

Cephalohematoma, subarachnoid hemorrhage, and intraventricular hemorrhage observed at autopsy may be attributed to hypoxic-ischemic injury. The definitive cause of death was most probably due to cardiovascular pathology resulting in a poor circulatory supply to the lungs.

This case of conjoint twins as presented by the authors will favor the fission theory as the separation seems to have occurred only in the cranial part with no involvement of the caudal part.

Conclusion

This report emphasizes anatomical presentations of conjoined twins with detailed morphological examination with emphasis on the cardiovascular malformations seen. This will allow for proper classification of conjoined twins and possibly guide future attempts at separation for better survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgments

The article was presented as a poster abstract presentation at the 29th scientific congress of the European Society of Pathology (ESP). Amsterdam 2017.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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