Turk Neurosurg 36(3):349-359, 2026
DOI: 10.5137/1019-5149.JTN.47617-24.5

Systematic Review

Received: 03.10.2024
Accepted: 03.06.2025
Published Online: 22.05.2026

A Child with Three Legs or Conjoint Parasitic Twin?

Gulyara CIGDEM[1], Mehmet Emin BOLEKEN[2]

[1]Harran University, Faculty of Medicine, Department of Neurosurgery, Sanliurfa, Türkiye
[2]Harran University, Faculty of Medicine, Department of Pediatric Surgery, Sanliurfa, Türkiye

This study has been presented by Çiğdem, G. and Boleken, M.E. under the title "Dublication of the Vertebral Column and Conjoined Twins: A Rare Type of Rachipagus" at the Ninth World Congress of Spine Surgery, held between November 3–5, 2022 in Athens, Greece, and published in a special edition of World Neurosurgery (OP-31, 28;2022).

Corresponding author: Gulyara CIGDEM ✉ gulyara@hotmail.com

ABSTRACT

AIM: Rachiphagus is a rare congenital anomaly in which conjoined twins are fused at the midline of the vertebral column region. When one twin is malformed, the condition is referred to as a parasitic twin. The term "parasitic twin" also encompasses cases involving extra limbs or limb-like structures. Despite ongoing research, the underlying causes of this condition remain unknown.

MATERIAL and METHODS: This is a systematic review of the literature with a case report. A literature search was done in English language in PubMed and Semantic –Scholar from 1952 to 2023. All articles and cases with excess leg or mass which were attached at the back of the spine were reviewed and analysed.

RESULTS: A total of 65 cases with rachipagus anomaly were included in this study. Females 37 (56.9%) were affected more than males 28 (43.1%). The lower limbs were in 41 (63.1%) cases, followed by rudimental limbs and mass were in 15 (23.1%) cases, upper limbs were in 8 (12.3%) cases and rudimental upper and low limbs were in 1 (1.5%) case. In majority of cases, accessory limb or mass were attached at lumbosacral region, 27 (41.5%), followed by 12 (18.5%) cases at lumbar region, 7 (10.8%) were at thoracolumbar region, 5 (7.7%) were at sacral and other regions. More than half of the cases 34 (52%) were in Asian countries, followed by 24 (37%) cases in Africa.

CONCLUSION: Rachipagus parasitic twin is congenital abnormalities that develops during embryogenesis and exists at birth with structural deformities of the spine and additional limb or limb like mass. This article presents a novel rachipagus case and systematically reviews the relevant literature.

KEYWORDS: Conjoined twin, Parasitic twin, Polymelia, Rachipagus twin, Tripedus

ABBREVIATIONS: ASD: Atrial septal defect, **CT:** Computed tomography, **EMG:** Electromyography, **F:** Female, **HCP:** Hydrocephalus, **ICM:** Inner cell mass, **IONM:** Intraoperative neurophysiological monitoring, **LMMC:** Lipomyelomeningocele, **LL:** Lower limb, **MRI:** Magnetic resonance imaging, **M:** Male, **MMC:** Meningomyelocele, **NTD:** Neural tube defect, **SEP:** Somatosensory evoked potentials, **USG:** Ultrasonography, **UL:** Upper limb, **VSD:** Ventricular septal defect

INTRODUCTION

Rachipagus is a rare embryonic malformation characterized by a pair of conjoined or parasitic twins fused along from the midline of the vertebral column. When two twins are teratologically unequal, the larger or the normal one of these twins that supports and provides nutrients for the parasitic twin is known as the "autosite" twin. In most cases, parasitic twins manifest as an excess limb or limb-like mass protruding from the back along the vertebral column, rather than developing into a fully formed infant. The parasitic twin is attached to an autosite twin. In most cases, parasitic twins appear as an excess limb or limb like mass arising from the back of the vertebral column instead of completing himself

Gulyara CIGDEM : 0000-0002-4888-1172 Mehmet Emin BOLEKEN : 0000-0001-9006-8364

as an infant. Whether this anomaly results from an improper separation of twins or is a manifestation of aneural tube defect (NTD) is still debated. Several hypotheses have been proposed for the etiology of rachipagus. Two main theories are associated with this, one of which is neural tube defects and the other is the aberrant embryogenesis of conjoined twins, but actual cause is still unknown (30,35,37,39,55,59,69). The incidence rate is 1 in 50,000 births and up to 1 in 1-2 million live births and seems higher in females than in males with a ratio of 3:1 (37,44). The first report was in 1802 by Vincenzo Malacarne who described it as a malformation with excess limbs in a body with the term polymelia (26). Accessory limbs on the back as a rachipagus were described by Deslongchamps in 1851 and by Jones in 1889 (17,24). A PubMed search revealed 26 articles on rachipagus since 1952, two articles on tripedus since 2009 and multiple articles on parasitic twins. But only 1 case of rachipagus was reported in 1991 with three fully developed normally functioning legs (13). Notably, only one case of rachipagus with three fully developed, normally functioning legs was reported in 1991 (13).

This report presents the second documented case of a patient with three fully developed, normally functioning legs, accompanied by a comprehensive systematic review of previously published cases.

MATERIAL and METHODS

The study was approved by the local ethics committee of Harran University (No:21; Date: 31.10.2022). Written informed consent was obtained from the patient's family for publishing this clinical report.

This is a report of a case with a third additional fully formed functioning leg with a comprehensive review of the literature and meta-analyses of the previously published cases with rachipagus. The study adhered to the PRISMA 2020 guidelines for systematic reviews. A literature search was conducted using PubMed and Semantic –Scholar, spanning 1952-2023 with key-words: including rachipagus, parasitic twins, tripedus, notomelia, and polymelia. The search was restricted to articles published in English. All articles and cases were reviewed for data, type and level of the excess limbs, autosite anomalies and surgical procedures. Only conjoined twins who were fused dorsally at the midline of the spinal column were included to this study. The cases with parasitic twining from other anatomical regions of the body (heteropagus, craniopagus, omphalopagus and others) and animal studies were excluded. Finally, 64 cases with rachipagus from 47 articles were reviewed. All data of the included cases were analysed by IBM SPSS statistic version 20. Student's t-tests were used for between group comparisons and significant of p value was accepted ($p<0.05$).

Present Case

A 1-day-old, full-term female new-born, weighing 3.00 kg, was referred to the Harran University hospital, Türkiye, in July 2022, with three fully formed functioning legs. There was no relevant family history and the mother had a normal pregnancy, with regular prenatal follow-ups. The tripedus condition was diagnosed in the third trimester, and delivery was via caesarean section, but prenatal ultrasonography (USG) images were not available.

Physical examination revealed a third limb attached to the baby's back at the midline of the lumbosacral vertebral column via an iliac bone. The third leg moved in conjunction with the other two legs or spontaneously (as confirmed by review of the video material). A femoral pulse was palpable on the medial side of the thigh of the third leg. A normal female perineum was present between the normal right and left legs, and the anus was present under the third limb (Figure 1). Rudimentary external genitalia were represented by a papilla anteriorly. All three legs were normally formed with a full range of movements and responded equally to painful stimuli.

X-ray and 3D-computed tomography (CT) of the pelvis demonstrated that the additional ileum articulating with the margin of the sacrum had a partial acetabular joint with a normally sized femoral head (Figure 2A, B). Magnetic resonance imaging (MRI) revealed a hip joint, femur, and thigh muscles in the third leg, as well as a posteriorly protruding spinal cord at the S2–3 level, along with rudimentary bowel loop, uterus, and vesicle sacs (Figure 3A, B).

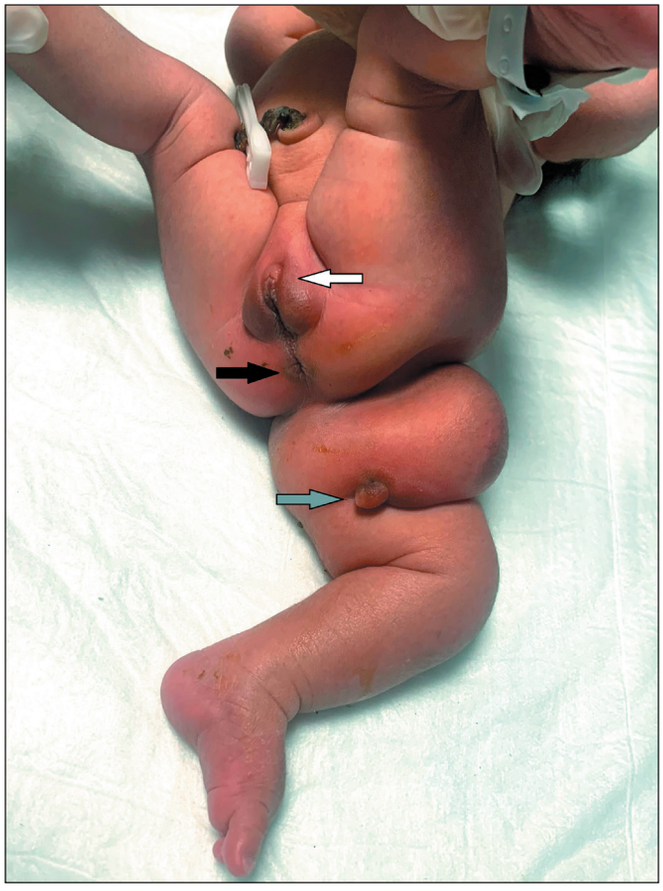

Figure 1: Lumbosacral attachment of a third leg to the back, featuring false external genitalia (black arrow). The female perineum is visible between the normal right and left legs (white arrow), and the anus is located under the third limb (grey arrow).

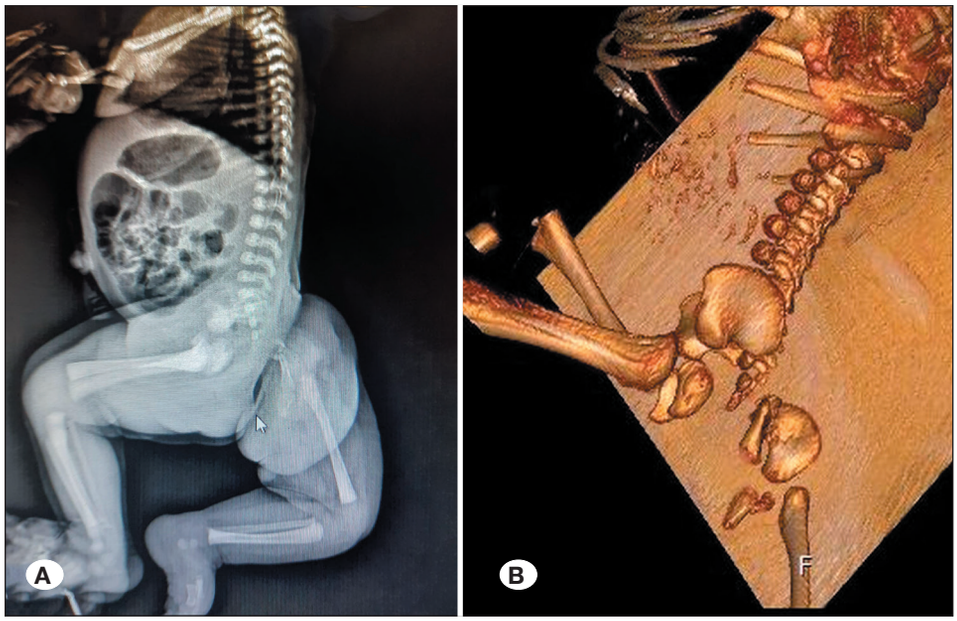

Figure 2: X-ray **(A)** and CT **(B)** lateral views of the pelvis, showing the additional ileum forming a partial acetabular joint with the head of the third femur and articulating with the sacral margin.

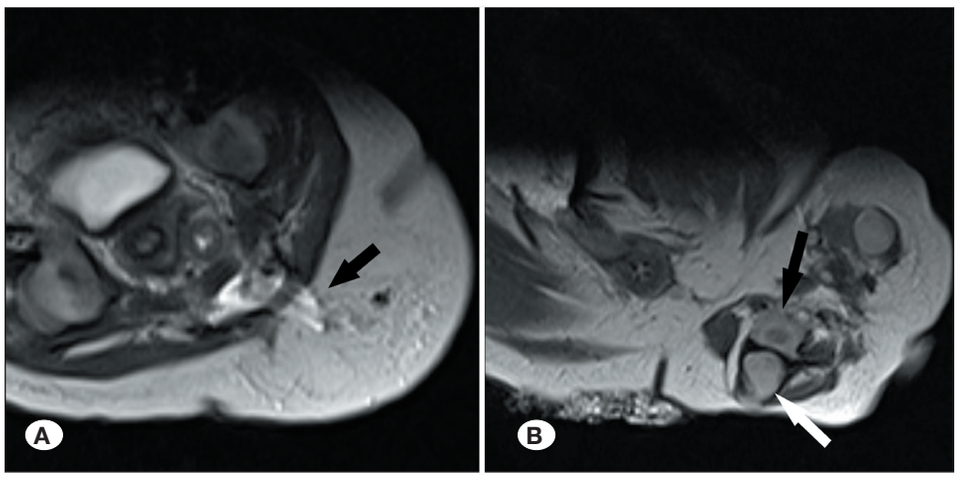

Figure 3: A) MRI-T2 weighted axial sections showing spinal cord roots protruding posteriorly at the S2–3 vertebral level (black arrow); **B)** A rudimentary bowel loop (white arrow) and uterus and vesicle sacs (black arrow) are visible.

Following a comprehensive assessment of all diagnostic tests, including CT and MRI scans, a surgical procedure was performed by a multidisciplinary team consisting of a pediatric surgeon and a neurosurgeon. The amputation was conducted under intraoperative neurophysiological monitoring (IONM) in order to precisely identify the location of neural structures and prevent any damage to the nerves of the first and the second legs. During the surgery, exploration was done while preserving an adequate flap for skin closure. The well-formed iliac bone of the parasitic leg was disarticulated from the autosite sacrum. Peripheral vascular bundles supplying the parasitic leg were ligated. The neuronal roots of the third lower limb were dissected under somatosensory evoked potential (SEP) monitoring. The SEP type of IONM was chosen because of its ease of application, ability to provide continuous monitoring during long surgeries, safety in infants, and compatibility with other monitoring techniques like electromyography (EMG) (Figure 4). The third leg was completely resected without damaging the neural structures of the sacrum, spinal cord, or both legs of the main body. The muscular cutaneous flap was well-performed, and the patient recovered smoothly, ultimately being discharged without any further complications.

RESULTS

Each case involving rachipagus was meticulously analysed to assess the data quality, excess limb characteristics, autosite anomalies, and surgical interventions (Table I).

The study found that rachipagus was more common in female infants, accounting for 56.9% (n=37), compared to male infants at 43.1% (n=28). Lower limbs were most commonly associated with rachipagus, occurring in 63.1% of cases (n=41), followed by rudimentary limbs and mases in 23.1% (n=15). Upper limbs were seen in 12.3% of cases (n=8), and a single case (1.5%) involved both rudimentary upper and lower limbs attached to the back of the spine. The findings are summarized in Table II.

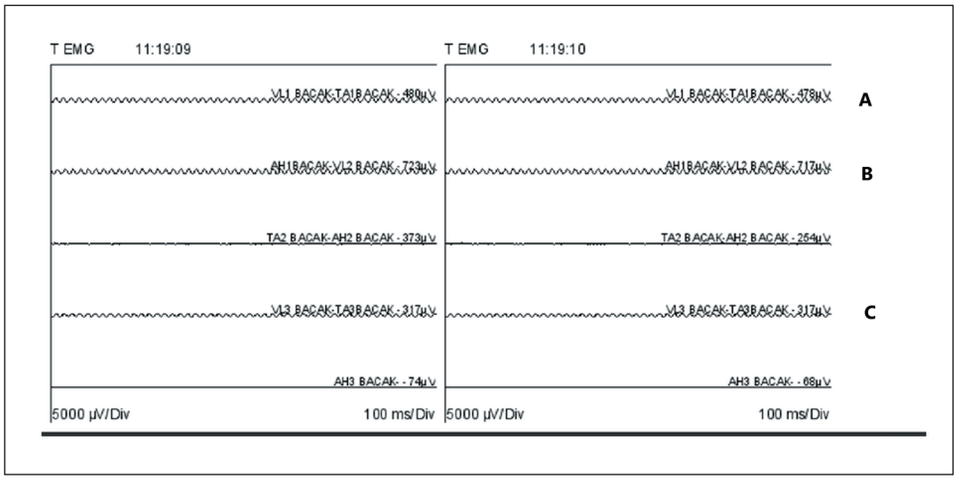

Figure 4: Intraoperative neurophysiological somatosensory evoked potentials monitoring of all three legs: **A:** right leg, **B:** left leg, **C:** additional third leg.

Table I: Summary of Cases of Rachipagus

Case	Age	Sex	Localization of appendage	Contents of parasite	Abnormalities in Autosite	Country
Copley and Derwael, 1991 (13)	8-days	M	Sacrum	Well developed LL	Spina bifida, MMC	South Africa
Ratan et al., 2004 (51)	2-days	F	Thoracolumbar	Vertebrae×2 Bowel Pelvis Bladder Fallopian tissue Limb with fused feet	Spinal dysraphism Tethered cord	India
Chadha et al., 2006 (12)	1-day	F	Lumbosacral	Anal dimple Digit like structures	Spinal dysraphism	India
Zhao et al., 2006 (72)	4-years	M	Lumbosacral	LL, pseudo navel, pseudo-penis	Hemivertebrae, Scoliosis, Lumbosacral vertebral dysplasia, Dislocation of the left hip	China
Amirjamshidi et al., 2006 (4)	6-weeks	F	Lumbar	Mass with thumb and fingers with nail beds	Spina bifida	Iran
Lende et al., 2007 (32)	2-years	F	Lower thoraco-lumbar	LL	Spina bifida MMC	Ethiopia
Snelling et al., 2008 (61)	4-months	M	Lumbar	UL like structures	High arched palate Retrognathia Posteriorly set ears Simian crease Spinal dysraphism Meningocele, hydromyelia Split, tethered cord	
Albert et al., 2008 (2)	9-months	F	Lumbosacral	Segmeted UL like structures	Spinal dysraphism Myelocystocele Teratoma	USA
Khan et al., 2009 (27)	1-day	M	Lumbosacral	LL, anal dimple, scrotum	Spina bifida	India
Sanoussi, 2010 (58)	3-months	M	Lumbar	LL	Lipoma, meningocele	Niger
	18-months	M	Thoracal	UL with two phalanges	Spina bifda	

Table I: Cont.

Case	Age	Sex	Localization of appendage	Contents of parasite	Abnormalities in Autosite	Country
Zhang et al., 2011 (71)	17-years	F	Thoracal	Breast, fallopian tube, bone	Spina bifida, Diplomyelia Scoliosis Tethered cord VSD	China
Debnath and Biswas, 2011 (15)				Well-formed limb, Intestine		India
Solak et al., 2012 (62)	2-months	F	Thoracal	A malformed UL	Lipoma	Türkiye
Ringo et al., 2012 (53)	3-days	M	Lumbosacral	LL, Phallus	MMC	Tanzania
Kota et al., 2012 (29)	5-months	M	Thoracal	Rudimentary limb	MMC	India
	3-months	M	Gluteal	Limb-like structures	MMC	
	1-day	F	Gluteal	Rudimentary limb	MMC, Anorectal anomaly, Duplication of external genitalia	
Kumar and Kumar, 2013 (31)	1-year	M	Thoracolumbar	Mass, fngers, phallus	MMC, Chiari malformation	India
Pandey et al., 2013 (45)	3-days	F	Lumbosacral	UL	MMC, Monoparesis	India
Murphy et al., 2013 (40)	7-months	M	Lumbosacral	Rudimentary LL	LMMC	USA
Oduor and Nyamal, 2014 (43)	9-days	M	Thoracolumbar	LL, UL, phallus	Spina bifida, club foot	Kenya
Bayri et al., 2014 (6)	1-day	M	Lumbosacral	LL, rudimentary pelvis	Meningocele	Türkiye
Parks and Mugamba, 2014 (47)	6-days	F	Thoracolumbar	UL, lactating breast	MMC, Kyphosis, Paraplegia, HCP, club foot	Uganda
Kim et al., 2014 (46)	1-day	F	Sacral	LL	Imperforate anus	Korea
Navaei et al., 2015 (42)	10-days	F	Lumbar	LL, hemipelvis, two toes	MMC, tethered cord	Iran
	5-days	M	Lumbar	Scrotum, primitive phallus	MMC, neurogenic bladder	
Nadeem et al., 2016 (41)	12-months	M	Lumbosacral	Limb, phallus, scrotum	Spina bifida	Pakistan
	8-months	F	Lumbosacral	Limb	Spina bifida	
	6-months	F	Lumbosacral	Limb	LMMC	
	4.5-months	F	Lumbosacral	Limb, bowel	Spina bifida, left leg weakness, urinary	
	1.5-months	F	Lumbosacral	Limb	incontinence Spina bifida	
Retnam et al., 2016 (52)	4-months	M	Lumbosacral	Rudimentary limb	MMC, Tethered cord	India
Awad et al., 2016 (5)	1-month	F	Lumbar	LL-foot, leg, knee, thigh, part of pelvis	LMMC	Egypt
Samy and Samin, 2016 (57)	28-days	F	Lumbosacral	LL, part of pelvis, phallus	Hemangioma, meningocele	Egypt
Sahlu et al., 2016 (56)	7-months	F	Lumbar	Mass, part of LL, phallus	LMMC Tethered cord Syringohydromyelia	Ethiopia
Mohammed et al., 2017 (38)	3-weeks	M	Cervicohtoracal	UL	Spina bifda	Nigeria

Table I: Cont.

Case	Age	Sex	Localization of appendage	Contents of parasite	Abnormalities in Autosite	Country
Kelani et al., 2017 (26)	5-days	M	Lumbar	LL, phallus	Meningocele Dermal sinus Umbilical herni	Niger
Raheja and Muhapatra, 2017 (50)	2-years	M	Lumbar	2 rudimentary limbs Anal dimple		India
Bodeliwala et al., 2017 (10)	1.5-year	F	Lumbar	LL, foot with syndactyled toes and nails	Club foot	India
Solomon et al., 2018 (63)		M	Lumbar	Mass, rudimentary bone	Meningocele	Ethiopia
Dejene et al., 2013 (16)	32-hours	F	Lumbosacral	LL, buttock, phallus	Cleft lip/palate, club foot	Ethiopia
2015	62-hours	M	Lumbosacral	Mass, phallus, scrotum	MMC, Cardiac (ASD, PDA)	
2018	18-hours	F	Lumbosacral	2 LL, phallus, pelvic bone, bowel	Rocker bottom feet	
Priyawansha et al., 2018 (49)	22-days	M	Thoracolumbar	LL	MMC, Umbilical hernia, Club foot	Sri Lanka
Khavanin et al., 2018 (28)	9-months	F	Cervicothoracal	Bilateral LL, pelvis, scapula	Diplomyelia, Lipoma, Thoracic kidney	USA
Adhikari et al., 2018 (1)	7-months (2-year)	M	Thoracal	LL	MMC, Diastematomyelia, Tethered cord, Hemicord	Nepal
Shrikesh et al., 2018 (60)	1-month	F	Lumbosacral	Rudimentary LL and genitalia	Large ASD	India
	3-days	F	Lumbosacral	Rudimentary LL		
Saaiq, 2020 (55)	5-months	F	Lumbosacral	LL, bowel	MMC, Tethered cord, Monoparesis	Pakistan
Zewdie, 2020 (70)	1-month	F	Lumbosacral	Mass with fngers	Meningocele	Ethiopia
	8-months	M	Lumbosacral	Mass with arm, hand Lung, lymph node	MMC, Tethered cord	
	2-months	F	Lumbosacral	Rudimentary limb, phallus	Meningocele	
	1.5-months	F	Thoracolumbar	Bilateral LL, phallus, part of pelvis,bladder	Myelocystocele, lipoma tethered cord, neurogenic bladder	
Pati et al., 2020 (48)	1-day	F	Cervical	2 accessory limbs	Cervical MMC,	India
	6-months	F	Lumbosacral	Rudimentary LL, phallus	Posterior cranial fossa, Mesocardia, ASD,VSD, Left to right shunting LMC, Tethered cord	
Mohindra et al., 2021 (39)	3-years	M	Lumbar	LL, phallus	MMC, Tethered cord	India
Bikoroti et al., 2021 (7)	1-day	F	Sacrococcygeal	Scalp, skull, cervical spine bone, and brain		Congo
Zhi, 2022 (73)	23-days	M	Thoracal	Penis and scrotum like mass		China
	6-months	F	Sacral	Hypoplastic LL with 7 toes,		
Djibrine, 2022 (18)	2-years	F	Thoracal paravertebral	Fatty clumps, cartilaginous tissue		Chad
	4-days	F	Sacral	LL		

Table I: Cont.

Case	Age	Sex	Localization of appendage	Contents of parasite	Abnormalities in Autosite	Country
Mathur et al., 2022 (35)	1-day	M	Sacrococcygeal	LL with 3 toes	Split notochord with MMC	India
	1-day	M	Sacral	Hypoplastic limb with 2 phalanges, rudimentary phallus	Ectopic right kidney, MMC	
Hailu et al., 2022 (22)	4-days	M	Lumbosacral	LL	Hemivertebra, LMMC	Africa
Present case	1-day	F	Lumbosacral	Full developed lower limb	Rudimentary loop bowel, Uterus, Vesical sacs	Türkiye
Total, n=65		M-28 F-37				

***F:** Female, **M:** male, **LL:** lower limb, **UL:** upper limb, **LMMC:** lipomyelomeningocele, **MMC:** meningomyelocele, **HCP:** hydrocephalus, **VSD:** ventricular septal defect, **ASD:** atrial septal defect.*

Table II: Distribution of the Cases in the Study

	Frequency (n)	Percent (%)
Sex		
Female	37	56.9
Male	28	43.1
Accessory Limb/Mass		
UL	8	12.3
LL	41	63.1
Rudimental limb and mass	15	23.1
UL+LL	1	1.5

***UL:** upper limb, **LL:** lower limb.*

The study showed that the lumbosacral region was the most common site of attachment of the accessory limb or limb-like mass in rachipagus cases, occurring in 41.5% of cases (n=27), followed by the lumbar region in 18.5% of cases (n=12). Other regions, such as the thoracolumbar region (10.8%) and sacral region (7.7%), had lower frequencies of attachment. The level of attachment in rachipagus cases with an accessory limb or mass is depicted in Figure 5.

Comparison of the levels of attachment of the excess limb/mass between males and females revealed no significant sex-based differences (Student's *t*-test, p>0.05).

The autosites in most cases presented congenital anomalies such as meningomyelocele, diastematomyelia, spina bifida, tethered cord, anorectal malformations, ectopic intestinal loops, neurogenic bladder, and rudimentary external genitalia.

34 (52%) cases with accessory limb or rachipagus were in Asian countries: India, Pakistan and others. 24 (37%) cases in Africa: Nigeria, Egypt, Ethiopia. 4 (6%) cases were in USA. 3 (5%) cases in Europe: all of them were in Türkiye (Figure 6).

DISCUSSION

Interest to congenital disabilities of humans or animals named as monsters appears since old ages and some of the cases are still exhibited in museums. Vallisneri in 1721 after observation of several cases of conjoined twins explained the formation of this condition as "from two germs or mature eggs, which by matching closely, over time attack and interpenetrate, so that they compose a doubled body… or with multiplied limbs, are born" (34). The progress in the number of the records with rachipagus for the last decades increased the interest in this malformation. However, its rarity limits opportunities for observation and hinders a full understanding of its embryological etiology and pathogenesis. This malformation represents a complex interplay of embryological and anatomical features. Several theories attempt to explain the mechanism for the development of an excess limb: 1) spontaneous incidence with a frequency of 10.25/1 000 000 births (25). 2) Incomplete twinning of monozygotic, monochorionic, and isosexual twins in the vertebral region associated with an additional limb or limb like mass and other organs or 3) NTDs resulting from aberrant neural fold formation during neurulation (11,26).

Kaufman, and Boer et al. suggest the first scientific fission and fusion theories of conjoined twinning. The fission theory is explained as an incomplete splitting of the embryo after fertilization on 13-15 days (11,25). The fusion theory posits the fusion of two separated embryonic disks around 13 days after fertilization. It means that two embryoblasts of monozygotic, monochorionic twins, instead of splitting away, merge together (11).

Grondahl et al. recorded the earliest embryogenesis of conjoined twins from a single zygote to an expanded blastocyst (21). Before implantation, the blastocyst contains two types of cells: the trophectoderm, which subsequently transform into extra-embryonic tissues and the inner cell mass (ICM), which is a pluripotent stem cell for embryonic tissues and transforms to all cell types of any organs of the embryo. The ICM can form two identical daughter cells through mitotic division (54,64,67). Failure of the ICM to separate in early embryogenesis may underlie the embryological theory of conjoined twinning (9).

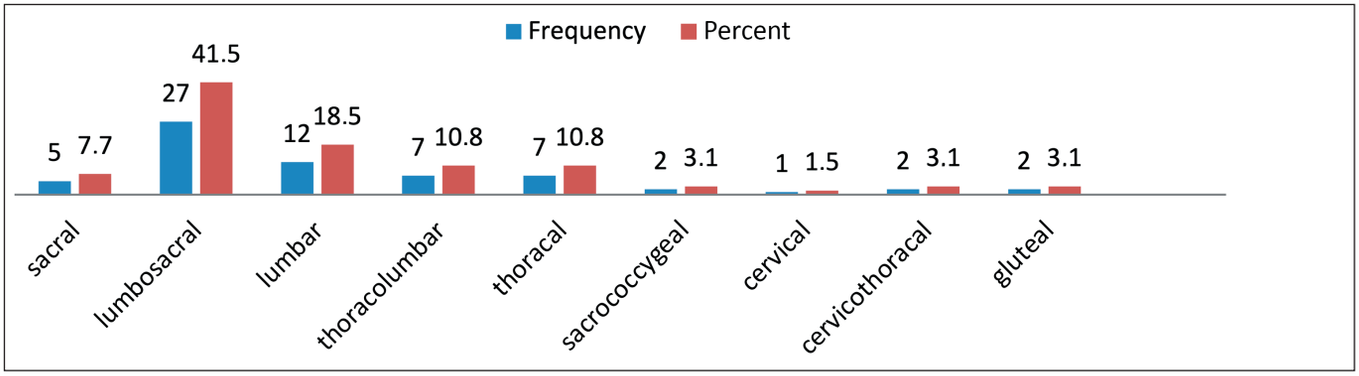

Figure 5: Level of attachment of the accessory limb/mass.

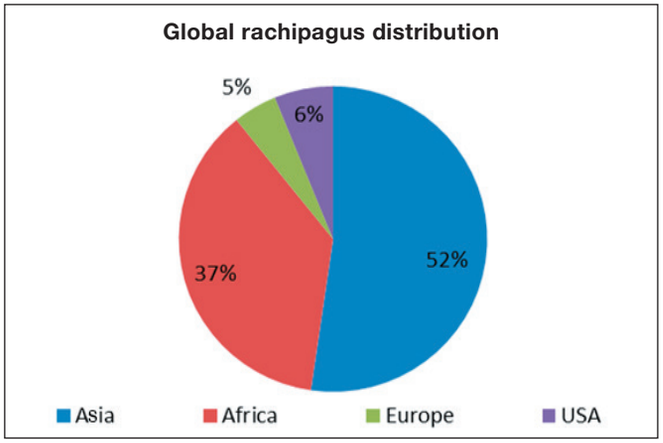

Figure 6: Global distribution of rachipagus cases.

Limbs develop between 4-5 weeks of embryonic development. Steinman collected data from pregnant women who gave birth to conjoined monozygotic twins following exposure to teratogenic factors during pregnancy. The results showed that pregnant women who used oral contraceptives and were exposed to teratogenic environmental factors had an increase in single egg duplication and conjoined twinning due to calcium depression and delayed implantation (36,65). According to Das and Mohanty-Hejmadi, vitamin A can cause polymelia by turning on or overexpressing limb-specific hox genes through unknown mechanisms (14). This implies a potential genetic cause, involving multiple genes responsible for limb development (23). Damage to the embryo during the 4–5 week stage can lead to limb developmental anomalies (68). This complex process can result in malformations, such as the parasitic twin occurring as an extra-limb or limb-like mass. Stephens et al. reported two cases of parasitic legs attached to the abdomen and suggested that ectopic legs may result from duplication of the Wolffian ridge. This duplication may more likely produce lower extremities than upper ones (66). According to our review (Table II), lower limbs are predominant, accounting for 41 cases (63.1%), often presenting as a parasitic mass.

Kelani et al. related rachipagus to NTDs as embryonic malformations characterized by an excess limb, classifying it as a dysraphic appendage (26). Most cases of rachipagus are linked to congenital NTDs, including MMC, LMMC, spina bifida, diplomyelia and tethered cord syndrome (26,31,45,71). Gardner also linked conjoined twins to neural tube closure defects, proposing that primary rupture of the neural tube can lead to secondary development of a limb-like mass or, in rare cases, a fully formed accessory limb containing skin, cartilage, bone, muscles, and nerves (20). Amirjamshidi considered congenital dorsal midline limb-like masses to be a type of hamartoma (3). According to Drews and Busch, based on his personal experience, parasitic conjoined twins can be viewed as extremely disorganized transitional forms, regardless of whether they manifest as parasites or teratomas (19). Each theory of rachipagus etiopathogenesis attempts to explain the formation of an excess limb, but every teratological, genetic, and embryological theory has its limitations.

The condition is more prevalent in females, with a female-to-male ratio of 3:1 (11). Our study highlights a significant disparity in the prevalence of additional limbs or masses between female and male infants, with females exhibiting a higher incidence. The distribution of limbs varied, with lower limbs being the most common, followed by rudimentary limbs and masses, upper limbs, and a rare occurrence of rudimentary upper and lower limbs located at the back of the spine. Globally, cases of the rachipagus are more frequently reported in Asia and Africa compared to European countries and America (11). Our study reflects this trend, with over half of the cases originating from Asian countries followed by African countries.

Conjoined twins are classified according to the site of attachment of the component of the parasitic twin: asymmetric or symmetric. The terminology describes the anatomical location, degree, and relationship of shared structures (25). These twins may share the spinal cord or have innervation from the same spinal cord, as observed in our case. Symmetric conjoined twins may be fused at ventral, dorsal, caudal, or lateral sites (11,37). Rachipagus twins are a type of symmetric conjoined twins characterized by dorsal fusion along the vertebral column above the sacrum (8). The fusion area may be at cervical, thoracal, or lumbar levels of the vertebral axis. Twins fused dorsally at the sacrum or coccyx are classified as pygopagus, distinct from rachipagus, which involves fusion higher up the vertebral column (3,8,37). Pypopagus twins often involve a parasite twin with a limb or limb-like

mass. Saaiq et al. described a morphologic classification for accessory limbs based on degree and differentiation: a) well-developed, b) moderately developed, c) mildly developed, d) poorly developed accessory lower limb (55). According to Saaiq's classification, our case is a rachipagus with a well-developed accessory lower limb with spinal dysraphism.

Modern imaging technologies like prenatal USG, MRI, or CT scans with 3D reconstruction enable early diagnosis of rachipagus anomalies (33). Surgical separation of conjoined twins poses significant challenges due to shared blood vessels, nerve roots, and vital organs, and potential cosmetic issues, necessitating meticulous pre-, intra-, and postoperative planning. Predicting the type of parasitic twinning, shared organs, neurological deficits, and associated malformations before surgery is crucial. A multidisciplinary team of neurosurgeons, pediatric surgeons, and plastic surgeons is essential for successful separation.

We have documented a rare case of rachpagus parasitic twinning with a fully developed additional low limb, exhibiting sensory and motor functions. Our case features a unique combination of anomalies, including three fully formed legs (tripedus), a protrusion and duplication of the spinal cord, with a rudimentary bowel loop, uterus, and vesicle sacs. The exact cause remains unclear, but multiple risk factors may contribute to the incidence of conjoined parasitic twinning during the early stage of fertilization.

The rarity of the rachipagus with a fully developed lower limb is a limitation of this study.

CONCLUSION

Rachipagus, conjoined or parasitic twins are a rare embryological anomaly characterized by dorsal fusion at the vertebral axis above the sacrum. The etiology of this anomaly is still unclear, but "fission or fusion" and erroneous of neural tube closure are the main theories of rachipagus. In most cases of parasitic twinning, the parasitic component typically consists of a lower limb with varying degrees of development, often attached to the lumbosacral region. A good prognosis can be achieved through early diagnosis, thorough preoperative examination, and meticulous microsurgery performed by a multidisciplinary team, incorporating spinal canal repair and cosmetic reconstructions.

Our case contributes to our understanding of rachipagus anomalies with NTDs. Further research, particularly human genetic DNA studies, is necessary to elucidate the embryogenesis of these congenital abnormalities.

Declarations

Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

Informed consent: Written informed consent was obtained from the parents/legal guardians.

AUTHORSHIP CONTRIBUTION

Study conception and design: GC
Data collection: GC
Analysis and interpretation of results: GC
Draft manuscript preparation: GC
Critical revision of the article: GC, MEB
Other (study supervision, fundings, materials, etc...): GC, MEB
All authors (GC, MEB) reviewed the results and approved the final version of the manuscript.

REFERENCES

1. Adhikari R, Thapa A, Pandey C: Rudimentary third arm arising from the upper back: a rare case. Grande Med J 1:41–43, 2019. https://doi.org/10.3126/gmj.v1i1.22405
2. Albert G, Campos M, Menezes A, Vogel T, Weinstein S: Rachipagus parasite associated with myelocystocele and diastematomyelia. Pediatr Neurosurg 44(5):418-421, 2008. https://doi.org/10.1159/000149912
3. Amirjamshidi A: Dorsal midline skin covered anomalies. A hamartoma or rachipagus? Childs Nerv Syst 32: 7–8, 2016. https://doi.org/10.1007/s00381-015-2927-8
4. Amirjamshidi A, Abbassioun K, Shirani Bidabadi M: Skin-covered midline spinal anomalies: a report of four rare cases with a discussion on their genesis and milestones in surgical management. Childs Nerv Syst 22: 460–465, 2006 https://doi.org/10.1007/s00381-005-0014-2
5. Awad T, Elsayem K, Elqazaz M: Dorsolumbar parasitic rachipagus twin: case report. Egypt Spine J 19:48–54, 2016. https://doi.org/10.21608/esj.2016.4013
6. Bayri Y, Tanrıkulu B, Ekşi M, Dağçınar A: Accessory lower limb associated with spina bifida: Case report. Childs Nerv Syst 30:2123-2126, 2014. https://doi.org/10.1007/s00381-014-2475-7
7. Bikoroti JB, Jahn A, MunezaS, M. Nyundo M, Gashegu JK, Buteera AM: A rare case of sacral parasitic rachipagus. Rwanda Med J 78(4): 12-16,2021. https://doi.org/10.4314/rmj.v78i4.2
8. Bindlish A, Sawal A: A detailed description and discussion on conjoined twins. Cureus 14(9):e29526, 2022. https://doi.org/10.7759/cureus.29526
9. Bishopa L, Jonesb B, Kelleyc D, Martin D: External parasitic twins. J Pediatr Surg Case Reports 40: 62-68, 2019. https://doi.org/10.1016/j.epsc.2018.08.015
10. Bodeliwala S, Singh D, Singh H, Iqbal M, Agarwal A, Khurana P: Spinal dysraphism with tripedus: A child with three legs. Neurol India 65(1):214-216, 2017. https://doi.org/10.4103/0028-3886.198209
11. Boer LL, Schepens-Franke AN, Oostra RJ: Two is a crowd: two is a crowd: on the enigmatic etiopathogenesis of conjoined twinning. Clin Anat 32(5):722-741, 2019. https://doi.org/10.1002/ca.23387
12. Chadha R, Lal P, Singh D, Sharma A, Choudhury SR: Lumbosacral parasitic rachipagus twin. J Pediatr Surg 41(1):e 45-48, 2006. https://doi.org/10.1016/j.jpedsurg.2005.10.070

13. Copley IB, Derwael EF: Investigation and treatment of a multiple limb birth. Br J Neurosurg 5: 281-287, 1991. https://doi.org/ 10.3109/02688699109005188
14. Das P, Mohanty-Hejmadi P: Vitamin A mediated limb deformities in the common Indian toad, Bufo melanostictus (Schneider). Indian J Exp Biol 38(3): 258-64, 2000.
15. Debnath B, Biswas SK: Heteropagus twinning on back - a case report. J Indian Med Assoc 109(7):514-515, 2011.
16. Dejene B, Negash S, Mammo T, Tadesse A, Getachew H, Miliard Derbew M: Heteropagus (parasitic) twins. J Pediatr Surg Case Rep 37:44–49, 2018. https://doi.org/10.1016/j.epsc.2018.07.019
17. Deslongchamps E: Memory on a monster double monomphalien of human origin, constituting a new genre called Rachipage. CR Séances Mem Soc Biol 3: 221-235, 1851
18. Djibrine MA, Ngaringuem O, Hagré YD, Adam A, Attimer K, Ouchemi Choua O: Heteropagus conjoined twins: five cases. J Pediatr Surg Case Rep 76:102086, 2022. https://doi.org/10.1016/j.epsc.2021.102086
19. Drews U, Busch C: Sacrococcygeal teratomas (SCTs): conjoined twins or conjoined daughter? Histopathology 54, 763–779, 2009. https://doi.org/10.1111/j.1365-2559.2009.03282.x
20. Gardner JW: Hypothesis: overdistension of the neural tube may cause anomalies of non-neural organs.Teratology 22: 229–238, 1980. https://doi.org/10.1002/tera.1420220212
21. Grondahl ML, Tharin JE, Maroun LL, Jørgensen FS: Conjoined twins after single blastocyst transfer: a case report including detailed time-lapse recording of the earliest embryogenesis, from zygote to expanded blastocyst. Hum Reprod 37(4):718-724, 2022. https://doi.org/10.1093/humrep/deac004
22. Hailu SS, Solomon DZ, Gebremedhin YG, Nour AS: Dorsolumbar parasitic twin associated with lipomyelomeningocele: a case report from a tertiary teaching hospital, Ethiopia, East Africa. Radiol Case Rep 17(10):3820-3824, 2022. https://doi.org/10.1016/j.radcr.2022.07.079
23. Hassanzadeh B, Rahemi A: Polymelia with unhealed navel in an Iranian indigenous young fowl. Veterinary Research Forum 8 (1): 85 – 87, 2017. PMID: 28473903. PMCID: PMC5413317.
24. Jones R, Larkin FC: Removal of accessory limb and meningocele from the back of a child, and its anatomy. Br Med J 2(1493):310–311, 1889. https://doi.org/10.1136/bmj.2.1493.310
25. Kaufman MH: The embryology of conjoined twins. Childs Nerv Syst 20: 508–525, 2004. https://doi.org /10.1007/s00381-004-0985-4
26. Kelani AB, Moumouni H, Issa AW, Younsaa H, Fokou H, Sani R, Sanoussi S, Denholm LJ, Beever JE, Catala M: Notomelia and related neural tube defects in a baby born in Niger: case report and literature review. Childs Nerv Syst 33(3):529-534; 2017. https://doi.org/10.1007/s00381-017-3337-x
27. Khan RA, Wahab S, Ghani I: Tripedus: a form of rachipagus twinning. Congenit Anom (Kyoto) 49(4):280-281, 2009. https://doi.org/10.1111/j.1741-4520.2009.00243.x
28. Khavanin N, Ruge JR, Vicari FA, Belin EJ, Kellogg RG, Steinberg JP: Parasitic rachipagus conjoined twin: case report. J Neurosurg Pediatr 22(3):313-316, 2018. https://doi.org/10.3171/2018.3.PEDS1822
29. Kota R, Srirampur S, Kannaiyan L, Irfan GM, Sharab H, Rao S: Parasitic twinning—varied presentations. J Dr NTR Univ Health Sci 1(3):174, 2012. https://doi.org/10.4103/2277-8632.102445
30. Krishna A, Lal P: Accessory limbs associated with spina bifida - a second look. Pediatr Surg Int 15: 248-250, 1999. https://doi.org/10.1007/s003830050568
31. Kumar A, Kumar R: Accessory digit and rudimentary male external genitalia associated with spinal dysraphism: a rare case of dysraphic appendages. Indian J Neurosurg 2:101–102, 2013. https://doi.org/10.4103/2277-9167.110233
32. Lende G, Wendemu W, Mørk S, Wester K: A girl with spina bifida, an extra leg, and ectopic intestinal loops--a "foetus in foetu" or a whim of the neural crest? Acta Neurochir (Wien) 149(10):1071-1075, 2007. https://doi.org/10.1007/s00701-007-1258-2
33. Liu J, Liu Y, Xue Y, Guo Y: Prenatal ultrasound-based diagnosis of fetal OEIS complex associated with lower limb polymelia and cardiac, hepatic dysplasia: A case report. Clin Case Rep 7(11):2153-2155, 2019. https://doi.org/10.1002/ccr3.2330
34. Magno G, Boer LL, Oostra RJ, Zanatta A: The role of the University of Padua medical school in the study of conjoined twins between 18th and early 19th century. Am J Med Genet A 188(12):3423-3431, 2022. https://doi.org/10.1002/ajmg.a.62938
35. Mathur P, Sharma S, Mittal P, Yadav RK, Barolia D: Heteropagus twins: six cases with systematic review and embryological insights. Pediatr Surg Int 38(7):963-983, 2022. https://doi.org/10.1007/s00383-022-05135-w
36. Mendilcioglu I, Simsek M: Conjoined twins in a trichorionic quadruplet pregnancy after ovulation ınduction with clomiphene citrate. Fetal Diagn Ther 24 (1): 51–54, 2008. https://doi.org/10.1159/000132407
37. Mian A, Gabra NI, Sharma T, Topale N, Gielecki J, Tubbs RS, Loukas M. Conjoined twins: from conception to separation, a review. Clinic Anat 30:385–396, 2017. https://doi.org/10.1002/ca.22839
38. Mohammed D, Haruna G, Sadusi MM, Sule SA, Musa MA: Parasitic limb attached to the back: a rare case of rachipagus parasitic conjoined twinning. Int J Health Sci Res 7:410–413, 2017. Available from: https://www.ijhsr.org/IJHSR_Vol.7_Issue.5_May2017/62.pdf. Accessed June 19, 2023.
39. Mohindra S, Batish A, Patil NR, Tripathi M: Fully developed accessory lower limb with tethered cord: are these two anomalies related? Childs Nerv Syst 37:325–328, 2021. https://doi.org/10.1007/s00381-020-04616-4
40. Murphy RF, Cohen BH, Muhlbauer MS, Eubanks JS, Sawyer JR, Moisan A, Kelly DM: An accessory limb with lipomyelomeningocele in a male. Pediatr Surg Int 29:749–752, 2013. https://doi.org/10.1007/s00383-013-3269-9
41. Nadeem M, Ijaz L, Rehman L: Vestigial accessory limbs with spina bifida: Our 5 – year experience. Pak J Neurol Surg 20:187–191, 2016. https://doi.org/10.1007/s00383-022-05135-w
42. Navaei AA, Habibi Z, Moradi E, Nejat F: Parasitic rachipagus twins; report of two cases. Childs Nerv Syst 31(6):1001-1003, 2015. https://doi.org/10.1007/s00381-015-2664-z

43. Oduor P, Nyamal K: Parasitic rachipagus conjoined twins: surgical management and follow-up of a case in Kenya. Ann Afr Surg 11:54–57, 2014. https://www.ajol.info/index.php/aas/article/view/114668
44. Ozkan-Ulu H, Yilmaz Y, Sari FN, Altug N, Uras N, Dilmen U: An unusual case of heteropagus: autosite with a complex cardiac malformation. Pediatr Neonatol 52: 358-360, 2011. https://doi.org/10.1016/j.pedneo.2011.08.011
45. Pandey A, Singh S, Pandey J, Gupta V, Verma R: Lumbosacral parasitic twin associated with lipomeningomyelocele: a rare occurrence. Pediatr Neurosurg 49:110–112, 2013. https://doi.org/10.1159/000358096
46. Park KB, Kim YM, Park JY, Chung ML, Jung YJ, Nam SH: An accessory limb with an imperforate anus. Ann Surg Treat Res 87:213–216, 2014. https://doi.org/10.4174/astr.2014.87.4.213
47. Parks C, Mugamba J: Accessory limb with myelomeningocele: a rare case challenging previously held beliefs. Childs Nerv Syst 30(12):2127-2128, 2014. https://doi.org/10.1007/s00381-014-2452-1
48. Pati AB, Mahalik SK, Naik S, Das K: Heteropagus twins: a tale of two. Neurolog India, 68 (6): 1453-1455, 2020. https://doi.org/10.4103/0028-3886.304090
49. Priyawansha Y, Munasinghe K, Manike N: A rare case of heterophagus twinning. Sri Lanka J Child Health 47:279–281, 2018. https://doi.org/10.4038/sljch.v47i3.8555
50. Raheja A, Mahapatra AK: Rachipagus parasitic twin. Neurol India 65:1443–1444, 2017. https://doi.org/10.4103/0028-3886.217969
51. Ratan SK, Rattan KN, Magu S, Rohilla S, Purwar P, Mathur SK: Thoracolumbar rachipagus parasite. Pediatr Surg Int 20(4):298-300, 2004. https://doi.org/10.1007/s00383-003-1131-1
52. Retnam RK, Dhanalakshmi V, Juanita FE, GnanaMuthu JG: Polymelia associated with lipomyelomeningocele: a case report. Int J Anat Res 4(3):2813-2816, 2016. http://dx.doi.org/10.16965/ijar.2016.341
53. Ringo Y, Drake D, Sillo T, Lakhoo K: Parasitic twin within spina bifida. Afr J Paediatr Surg 9(3):240-242, 2012. https://doi.org/10.4103/0189-6725.104728
54. Rosenberg LE, Rosenberg DD: Human Genes and Genomes. Chapter 4 - Growth, fırst edition, Development and Reproduction, Elsevier 2012, 4: 27-50. ISBN: 9780123852120. eBook ISBN: 9780123852137
55. Saaiq M, Zimri FK, Zaman K: Successful treatment of well-developed accessory lower limb associated with spinal dysraphism: World J Plast Surg 9 (1): 73–81, 2020. https://doi.org/10.29252/wjps.9.1.73
56. Sahlu A, Mesfin B, Tirsit A, Debebe T, Wester K: Parasitic twin--a supernumerary limb associated with spinal malformations. A case report. Acta Neurochir 158(3):611-614, 2016. https://doi.org/10.1007/s00701-016-2710-y
57. Samy S, Amin H: Successful separation of rachipagus parasiticus neonate: case report and review of the literature. Egypt Spine J 20:51–60, 2016. https://doi.org/10.21608/esj.2016.4096
58. Sanoussi S, Rachid S, Sani CM, Mahamane B, Addo G: Rachipagus: a report of two cases - thoracic and lumbar. J Surg Tech Case Rep 2(1):27-29, 2010. https://doi.org/10.4103/2006-8808.63720
59. Sharma G, Mobin SS, Lypka M, Urata M: Heteropagus (parasitic) twins: a review J Pediatr Surg 45: 2454–2463, 2010. https://doi.org /10.1016/j.jpedsurg.2010.07.002
60. Shrikesh S, Singh US, Singh N, Sharanabasappa G, Amit N, Tanvir RK, Chetan S: Conjoined twins: a tertiary care centre experience. Sch J App Med Sci 6:1477–1480, 2018. https://doi.org /10.21276/sjams.2018.6.4.18
61. Snelling CM, Ellis PM, Smith RM, Rossiter JP: Lipomatous lumbar mass with an attached digit and associated split cord malformation. Can J Neurol Sci 35(2):250-254, 2008. https://doi.org /10.1017/s0317167100008738
62. Solak A, Ergün S, Polat I, Sahin N, Genç B: A rare form of heteropagus twinning: three-armed infant with spinal dysraphism. Case Rep Pediatr 2012: 831649, 2012. https://doi.org /10.1155/2012/831649
63. Solomon DZ, Fekadu N, Gorfu Y: Rachipagus parasitic twin in an Ethiopian neonate: a case report from the Horn of Africa. Int Res J Med Med Sci 6:56–59, 2018. https://doi.org/10.30918/IRJMMS.63.18.030
64. Spitz L: Conjoined twins. Br J Surg 83(8):1028-1030, 1996. https://doi.org/10.1002/bjs.1800830803
65. Steinman G: Mechanisms of twinning. V. Conjoined twins, stem cells and the calcium model. J Reprod Med 47(4):313-321, 2002. PMID: 12012884
66. Stephens TD, Siebert JR, Graham Jr JM, Beckwith JB: Parasitic conjoined twins, two cases, and their relation to limb morphogenesis. Teratology 26(2):115-121, 1982.https://doi.org/10.1002/tera.1420260203
67. Susana M, Lopes CS, Christine L. Mummery: Handbook Of Stem Cells 2:1-32, 2004.
68. Verma S, Khanna M, Tripathi VN, Yadav NC: Occurrence of polymelia in a female child. J Clin Imaging Sci 3:18, 2013. https://doi.org/10.4103/2156-7514.111235
69. Wilkes SL, Choi JJ, Rooks VJ: Lumbosacral lipomyelomeningocele with anomalous osseous limb in a 3-month-old female. Radiology Case Reports. Radiol Case Rep 10(1); 1051, 2015. https://doi.org/10.2484/rcr.v10i1.1051
70. Zewdie K, Negash S, Bizuneh Y, Woldemichael F, Temesgen F: Rachipagus parasitic twins: A case series and review of literature. Interdisciplinar Neurosurg 24-101049, 2021. https://doi.org/10.1016/j.inat.2020.101049
71. Zhang J, Duan H, Zhang Y, Yi Z, Bao S: Parasitic rachipagus conjoined twins with spina bifida, diplomyelia, scoliosis, tethered cord syndrome, and ventricular septal defect: case report. Neurol Med Chir 51(10):736–739, 2011. https://doi.org/10.2176/nmc.51.736
72. Zhao L, Li MQ, Sun XT, Ma ZS, Guo G, Huang YT: Congenital lumbosacral limb duplication: a case report. J Orthop Surg (Hong Kong) 14:187–191, 2006. https://doi.org/10.1177/230949900601400216
73. Zhi X, Hu B, Zhao X, Chen J, Gu C, Pu L, Fang Y, Cai C: A cohort of five cases with asymmetric conjoined twining and literature review. Pediatr Surg Int 38(1):169-181, 2022. https://doi.org/10.1007/s00383-021-05006-w