

## Rare Case of Conjoined Twins: Pictorial Essay

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### Abstract

Conjoint twins are rare, presenting a unique challenge to radiologist and pediatric surgeons. For surgical separations, accurate preoperative imaging is necessary for operative planning prognostic information as well as important for patient counseling. There are different types of anatomical connections. MRI and CT scan give excellent anatomical and bony details like organ position, shared organs and vascular anatomy. We present a pictorial essay on a previously unreported variation of pygopagus twins.

**Keywords:** Conjoint Twins, Pygopagus

### Introduction

Conjoined twins are rare, but the exact prevalence is unknown. The estimated prevalence in the literature varies widely, from 1:50,000 to 1:200,000.<sup>1,2,3,4</sup> An increased prevalence is observed in parts of Southeast Asia and Africa, with reported occurrence ranging from 1:14,000 to 1:25,000.<sup>5</sup> There is a female predominance on the order of 3:1.<sup>4</sup> The twins are monozygotic; mono amniotic mono chorionic; there is failure of separation of the embryonic plate between 13 and 17 days of gestation.<sup>3,6,7,8</sup> Conjoined twins are classified according to the most prominent site of

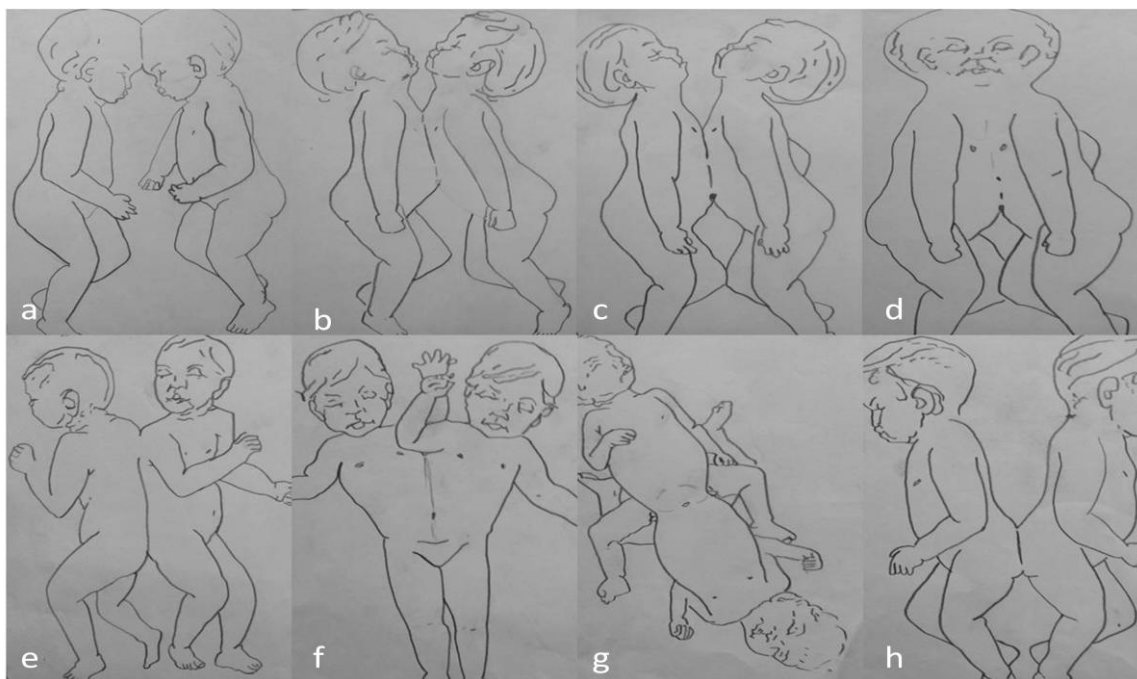
connection (Table.1): the thorax [thoracopagus], abdomen [omphalopagus], sacrum [pygopagus], pelvis [ischiopagus], skull [craniopagus], face [cephalopagus], or back [rachipagus].

Twin Type	Conjoined Area	Percentage of cases
<b>Thoracopagus</b>	Thorax	40
<b>Omphalopagus</b>	Abdomen	33
<b>Pygopagus</b>	Sacrum or buttocks	19
<b>Ischiopagus</b>	Pelvis	6
<b>Craniopagus</b>	Cranium	2
<b>Parapagus</b>	Ventro lateral Area	-

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### Imaging Importance

Conjoined twins present unique challenge to radiologists, obstetrician and pediatric surgeons. Planning of surgery is aided by accurate preoperative imaging. The area of



**Fig.1: a.Craniophagus b.Thoracopagus c.Omphalopagus d.Cephalopagus e.Rachipagus f.Parapagus g.Ishiopagus h.Pygopagus<sup>9</sup>**



**Fig. 2: Both Male babies of 2 month with fusion at lumbosacral region with separate GU System and Anus. On Skiagram there is fusion of babies at lumbo sacral region with non visualization of sacrum in baby on left.**

fusion largely determines the imaging modality used. Each set of conjoined twins is unique, an imaging strategy to accurately define anatomic fusion, vascular anomalies and other associated abnormalities is important for surgical planning and prognostic information.

### **Pygopagus [usual presentation]**

Pygopagus twins are joined dorsally, facing away from each other and sharing the sacro-coccygeal and perineal regions. Fusion of sacral vertebrae frequently occurs, but the spinal cords usually remain separate. Usually, there is a single anus with one or two rectums, the remainder of the intestine being separate. Twins may share a genitourinary system with a single bladder and urethra. The upper bodies are not fused, and there are four arms and four legs.<sup>2,9,13,14</sup>



**Fig. 4: On CT scan (Sag and axial) Spina bifida from L4 vertebra downwards with fusion of filum terminale.**

### In our case

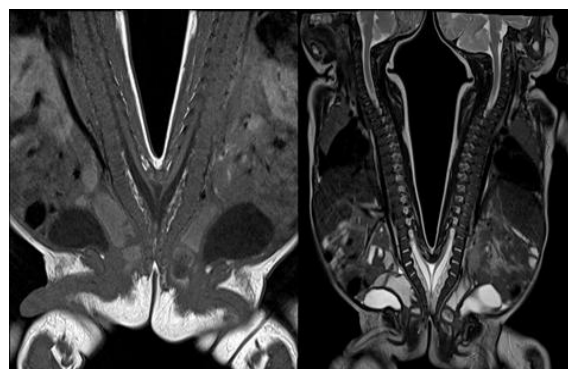
We present a variant of Pygopagus conjoined male twin, where the only area of fusion was filum terminale without any bony, vascular, genitourinary or other anatomic fusion. This is probably the: “First case of Pygopagus twins to present in such a manner.” Hence we highlight its imaging features on conventional radiography/CT/MRI as well as surgery. After taking clinical detail and morphology of conjoined twin, patient was subjected to digital radiography, then CT on Siemen definition- As 64 Slice MD CT scanner and 1.5T MRI Symphony TIM 18 channel MRI scanner.



**Fig.5: 3D rendering of the conjoined twins.**

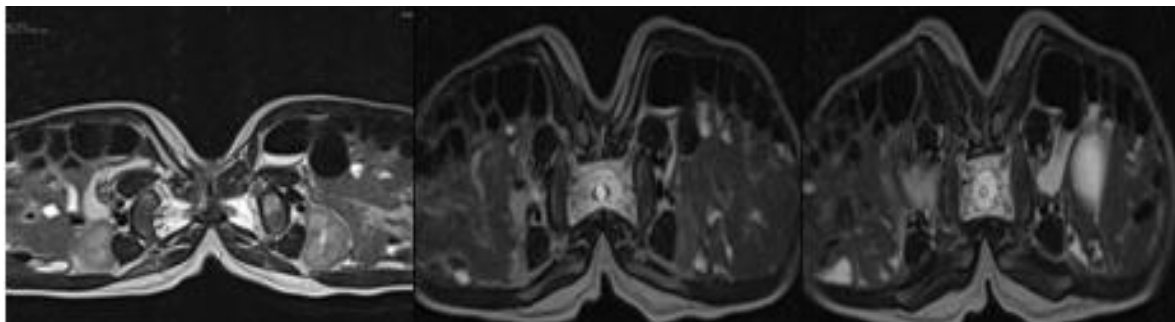
### Discussion

This pygopagus baby was delivered via cesarean section outside our institute and then referred for further management. These conjoined twins were evaluated at 2 months of age. During the preoperative planning, two separate surgical teams were established, each with responsibility for one of the twins; an experienced pediatric surgeon took charge for overall coordination. We had 2 pediatric surgeons, 2 neuro surgeons and 2 anesthetist in our team. Surgery was done successfully without complication as the surgical prognosis of this twin was better, where organs were not shared between twins like in usual presentation of pygopagus and omphalopagus twins described by Sanders RC et al.<sup>12</sup>



**Fig.6: On T1 Sag & T2 Sag WI: Both the cords are low lying and tethered and are seen lying approximately at L5-S1 level, thereafter the filum terminale are seen fused together at approx. S2-S3 level in single thecal sac.**

Preoperative imaging of conjoined twins and their interpretation presents and poses a big challenge to a radiologist. Their separation is also very challenging for the pediatric surgeons hence an imaging strategy to accurately define anatomic fusion, vascular anomalies, and associated abnormalities are important for proper surgical planning and prognostication.



**Fig.7: T2 Axial WI: Two Separate Cord Seen at upper lumbosacral region; Thereafter the filum terminale are seen fused together at approx. S2-S3 level in single thecal sac.**

With accurate preoperative imaging, rate of successful separation of conjoined twins has improved. The site of conjunction largely determines the imaging modalities used. MR imaging provides excellent overall anatomic demonstration, and visceral conjunction. Organ position is best assessed with this multiplanar technique. CT also provides good anatomic detail; CT which when performed with standard bone algorithms and in addition to three-dimensional reconstruction helps in excellent evaluation of bony fusion abnormalities and defects, and thus MR imaging and CT can be used together in conjunction to guide other imaging investigations.



**Fig. 8: Post Operative: Babies Successfully Separated**

### Conclusion

Certain imaging modalities and investigations are desirable in evaluating conjoined twins and can be determined according to the primary site of conjunction which highly varies. Thus even though each set of twins is unique; certain broad based generalizations can be made, but still exact imaging performed must be tailored to the twins involved and the requirements of the pediatric surgeons for conducting their separation.

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