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Dipygus
Complete Caudal Duplication
GROSS APPEARANCE
Complete Caudal Duplication & Gastroschisis
A full term baby boy 4 hours age.
* Have tetrapus (four legs) baby.
* Four lower limbs attached to double perineum, double pelvis.
* Double male external genitalia.
* Bilateral imperforated anus.
* Gastrochisis.
* The urine coming from left urethra stained with meconium.
Monocephalus Dibrachius Quadripus & Gastrochisis
INVESTIGATION
* X-ray shows segmentation disorder vertebral column started from thoracic part.

* Duplication of last lower 3 ribs in lifte side.

* Duple pelvis, hypoplastic medial half of pelvic bones with femur fracture in right medial lower limb.
The attachment of left pelvis to vertebral column is higher than the right pelvis and lateral to it.

* I.V.P shows 2 kidney but there is duplication in the renal pelvis and calyx of left one, 2 ureters, 2 urinary bladders.
Duple pelvis, hypoplastic medial half of pelvic bones
Duplications of the lower three ribs on the left side & segmentation of vertebrae from mid thoracic caudally
The attachment of left pelvis to vertebral column is higher than the right pelvis.
Duplication in the urinary bladder
Duplication in the L.t renal pelvis
FIRST OPERATION
* **Exploration** reveal single liver. Spleen, duodenum, jejunum.

* The right terminal ileum has Meckel’s Diverticulum.

- 2 cecum, 2 appendix, 2 colon, 2 kidneys from each one, one ureter ends in each urinary bladder.

* Two pelvis conjoined laterally.

* **Enteric cystostomy** done with primary closure of abdominal wall defect (Gastroschisis).
SECOND OPERATION
*Nine days later, 2nd operation done, amputation of additional central lower 2 limb with resection of hypoplastic corresponding 2 pelvis bones.

*Re approximating the pelvis by s. stile suture to form new pelvis followed by gallow’s traction for 2 weeks.
At 6 weeks age, the 3rd operation done:

* Resection of the enteric cyst and end to end ileoileal anastomosis
* Resection of Meckel's diverticulum
* Total colectomy of left colon
* Anoplasty for right side.
Patient Discharged home at 8 week old To be readmitted for The 4th operation at 6 M old (resect the left external genitalia and anastomose right and left urinary bladder)

Feedback
At 14 weeks he developed severe aspiration pneumonia and Septicemia, expired at 16 W old
Dipygus

Complete Caudal Duplication
Caudal Duplication Syndrome

Dipygus

A spectrum of an extremely rare congenital anomalies.

It is a form of *monocephalic twins* with *duplication of the caudal region* of an embryo.

Mostly due to *fission* of the germinal disc at the caudal end in the 2ed week of gestational age.
This *fission* is arrested and did not continue to the cephalic end.

Completely arrested development of the upper part of one twin.

It might be considered as a *parasite* type of conjoined twins specially if the duplicated part is not completely well developed (parasitic twin attached to the host twin)
Dipygus

* complete:
  somatic and visceral duplication of the lower body.

* partial:
  only visceral duplication
  or with one lower limb duplication.

2 Similar cases (complete caudal duplication)
  somatic and visceral duplication.

24 cases (partial caudal duplication).
Separation of Monovular Twins

Caudal duplication:
The fission is started from umbilical level to caudal end.

Cephalic duplication:
The fission started from umbilical level to cephalic end.

Twin's:
If it is complete fission, it will separate the germinal disc and it will lead to separated or conjoined twin's.
*According to the **site of conjunction:**

- Skull (**craniopagus**)  
- Thorax (**thoracopagus**)  
- Upper addo (**xiphopagus**)  
- Lower abdo (**omphalopagus**)  
- Pelvis (**ischiopagus**)  
- Sacrum (**pygopagus**)  

*The responsible **factors** might be:**

Teratogenic, Environmental or Genetic
Two cases of dipygus were reported in the English literature till date:

Francisco Lentini

Myrtle Corbin
parasite type of conjoined twins
Embryonic Development

- **Wnt (Wingless/integrated)-signaling**: responsible for the basic developmental processes. The Wnt family is composed of 19 glycoproteins.
- **Axis patterning**, **body axis formation**
- **Cell fate specification**
- **Cell proliferation**, **control of asymmetric cell division**
- **Cell migration**
Mutations in **WNT genes** or its pathway components lead to specific developmental defects of the cephalo-caudal axis especially the **notochord** urogenitals the **intestines**
The occurrence of caudal duplication is the result of disruption of regulation of WNT pathway at the early part of embryogenesis.
summary

Caudal Duplication

- Monocephalus
- Dibrachius
- Quadripus
- Twin

Urgent operation is indicated to correct other congenital anomaly that does endanger survival.

Correction can be carried out electively staged, preferably between 3-6 months.