

Newborn with Large Open Myelomeningocele Associated with Kyphoscoliosis: A Case Report



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Background

The incidence of spinal dysraphism has declined drastically due to the routine use of prenatal folate. With antenatal screening and termination of pregnancy, it is rare to see newborn with huge open myelomeningocele (MMC).

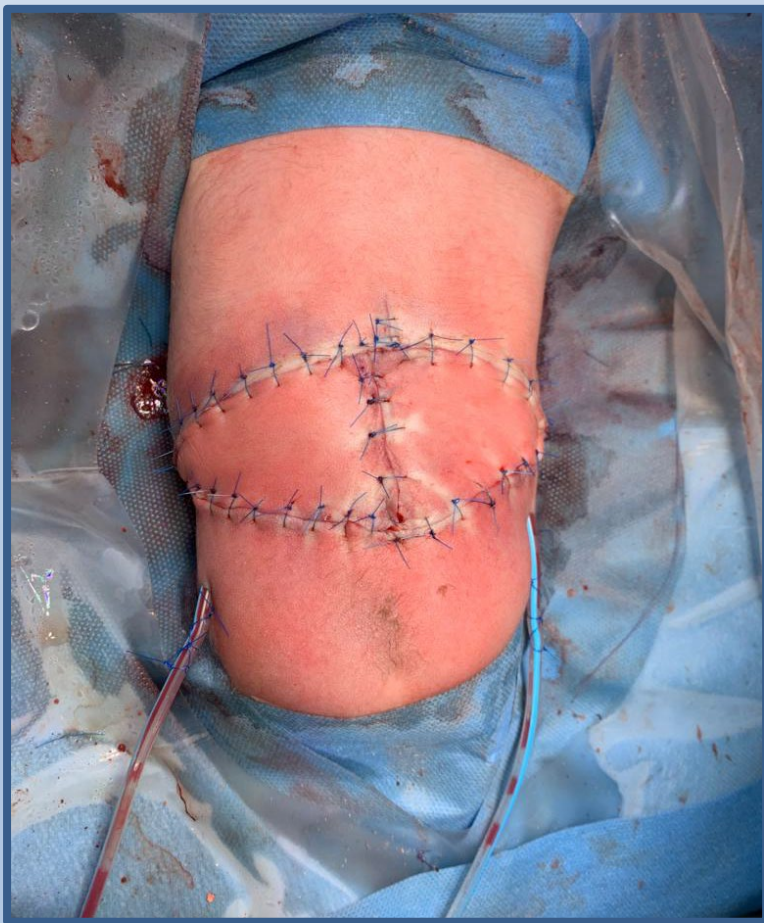
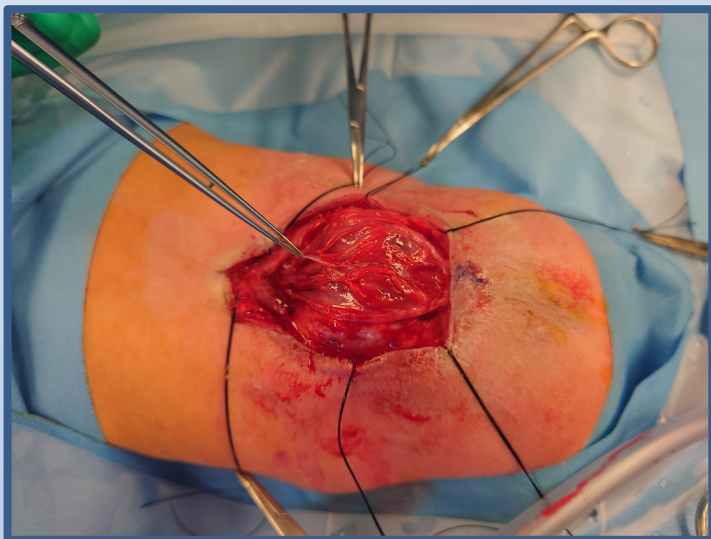
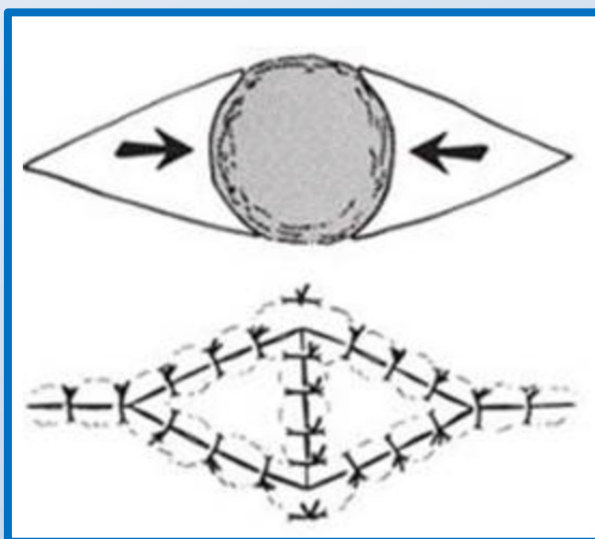
Case Summary

- A pregnant woman with substance abuse and drug-induced psychosis presented at 36 weeks of gestation without any previous antenatal checkup.
- Antenatal USG: open neural tube defect, hydrocephalus, deformed lower limbs with clubfoot, paraplegia
- Delivered by Caesarean section at 38+3 weeks
- Neonatal physical exam:
 - ① 5cm x 4cm open MMC with active CSF leakage
 - ② Severe kyphoscoliosis: marked kyphotic hump at L4
 - ③ Full anterior fontanelle (hydrocephalus secondary to Chiari II)
 - ④ Bilateral deformed lower limbs with clubfoot + paraplegia
 - ⑤ Lax anal tone



Immediate Management

- Immediate MMC repair
- Kyphectomy:
L3 cordectomy + L3-L5 partial discectomy & corpectomy
- Wound closure: V-Y advancement flap by Plastic team

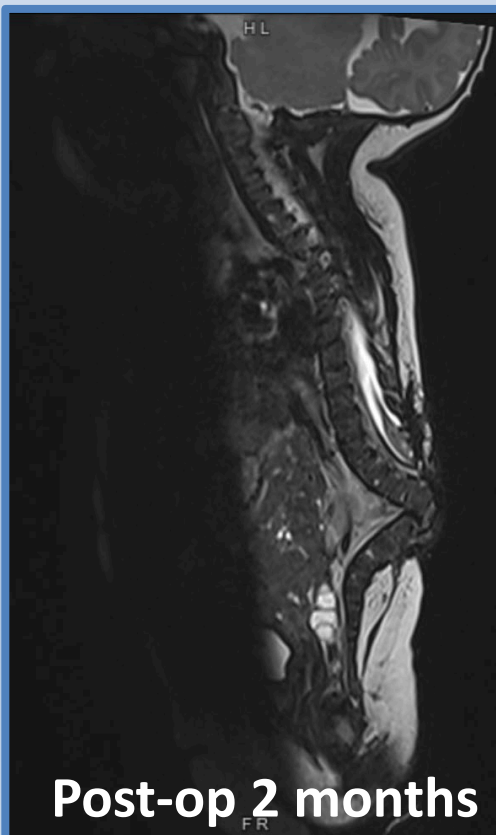
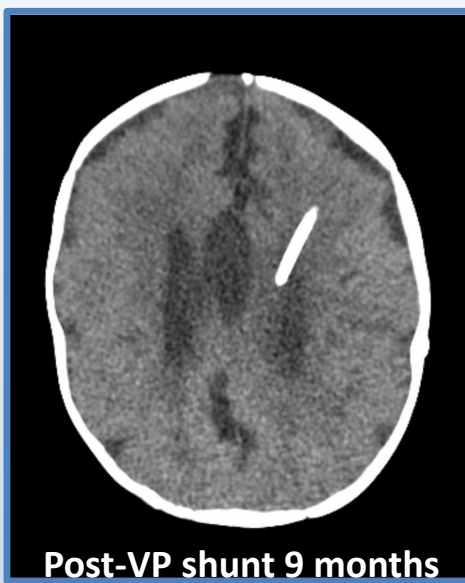


Outcomes

VP shunt insertion for hydrocephalus on post-kyphectomy day 13

At post-operative 1 year:

- Able to lie supine with good wound healing
- Good eye contact, smiling, started vocalization
- Good head control with age-appropriate fine motor skills
- Neurogenic bladder & fecal incontinence; on daily clean intermittent catheterization
- Ankle-foot orthoses for maintenance of foot shape



Discussion

- Neonatal kyphectomy is rarely required in MMC patients. Previous case series of neonatal kyphectomy have demonstrated association with better wound healing after MMC repair with excellent initial correction.^{1,2}
- After open MMC repair, close monitoring of head circumference and ventricular size is crucial for fear of acute hydrocephalus. VP shunt is required in up to 80% of patient.^{3,4}
- In our patient, intra-operative challenges included positioning, ethical judgement on sacrificing neural tissue for kyphectomy, prevention of CSF leak and closure of large skin defect.
- The decision of kyphectomy was made intraoperatively in view of severe kyphosis confirmed by intra-op X-ray, large skin defect, and the absence of caudal spinal cord function.



Conclusion

In neonates with MMC and kyphosis with complete loss of neurological function, kyphectomy in conjunction with MMC repair can be considered for early correction of kyphosis.

References

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