Two cases of very large heads…

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Case 1: MH   DOB 2/10/15

Consanginous parents, 1 previous well child
20wk scan: ventriculomegaly
22wk scan: macrocephaly, holoprosencephaly
“*Very poor prognosis discussed with [name], she is fully aware will not survive beyond the immediate neonatal period…options of karyotype and/or termination discussed and declined*”
26wk scan: “termination discussed again but declined” For prophylactic steroids

Case 1

Antenatal counselling

- Late call (within 1 week of projected delivery) to SHO to counsel
- Difficulties with setting up a joint counselling in a timely manner.

Case 1

Delivery / Postnatal course

Born at 33+2 weeks via c-section, paeds not present at delivery as planned.

Able to tolerate small bottle/cup feeds, remained with Mum rooming in

D3 – wish to be at home. ACP written. Not for intubation, not for compressions, PRN analgesia for comfort

Decision for vitamin K to be given, follow-up organised

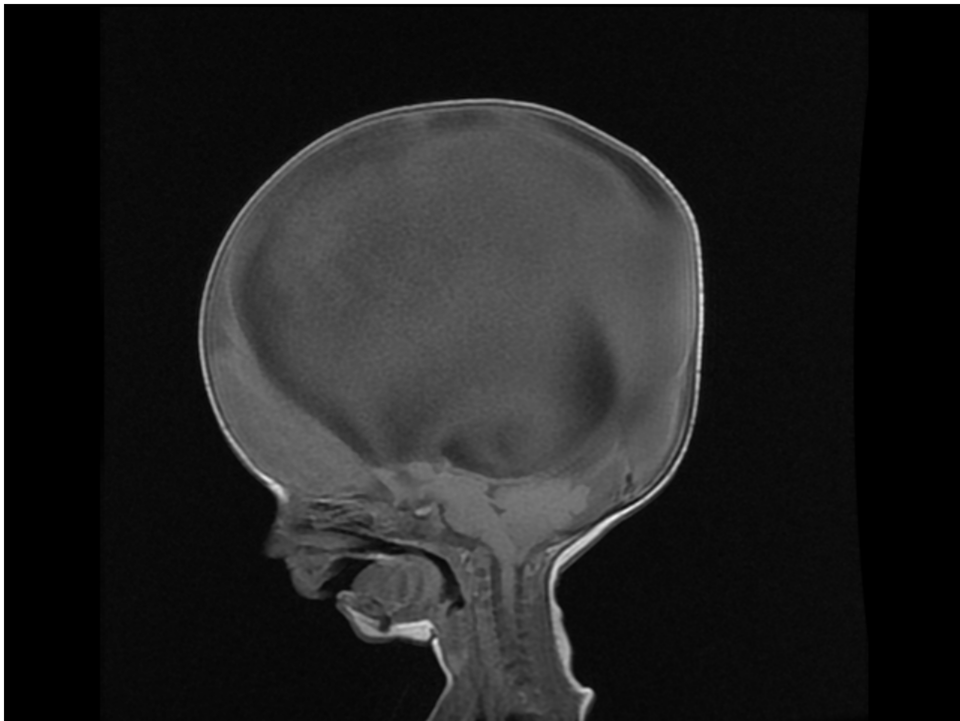
D/C home D4 with community nurse input

Case 1

What happened next?

- MRI at 4 weeks of life: Holoprosencephaly variant with large monoventricle
- Discussed with neurosurgery due to rapidly incr HC – urgent clinic review
- VP shunt inserted Nov ‘15 (8 weeks old) → infection, seizures, revision Feb ‘16, then removal due to repeated blockage

Case 1



Current Issues

6 months old…

- VP shunt removed, awaiting new

- Impaired hearing
- R cataract
- Focal seizures
- Likely ACTH deficiency
- Unsafe swallow – NG feeds. Weight maintained on 25th centile
- GDD

Case 1

Case 2: AP DOB 15/03/16

MCDA twins

AN scans for twin 2:

Severe bilateral ventriculomegaly (>15mm)
Poorly formed cerebellum
Small connection between intrahemispheric fissure

Case 2

Antenatal counselling

Obstetric + neonatal, discussed:

- Options for further investigation and intervention antenatally
- Risks to other twin
- Likely delivery plan
- Plan postnatally – for NICU to further assess and plan

This plan available to all team in high risk folder, and communicated in letters to parents

Case 2

Delivery / Admission to NICU

SOPL at 32+3 weeks – delivered by c-section due to ventriculomegaly + breech for both twins.

Minimal resus required at delivery

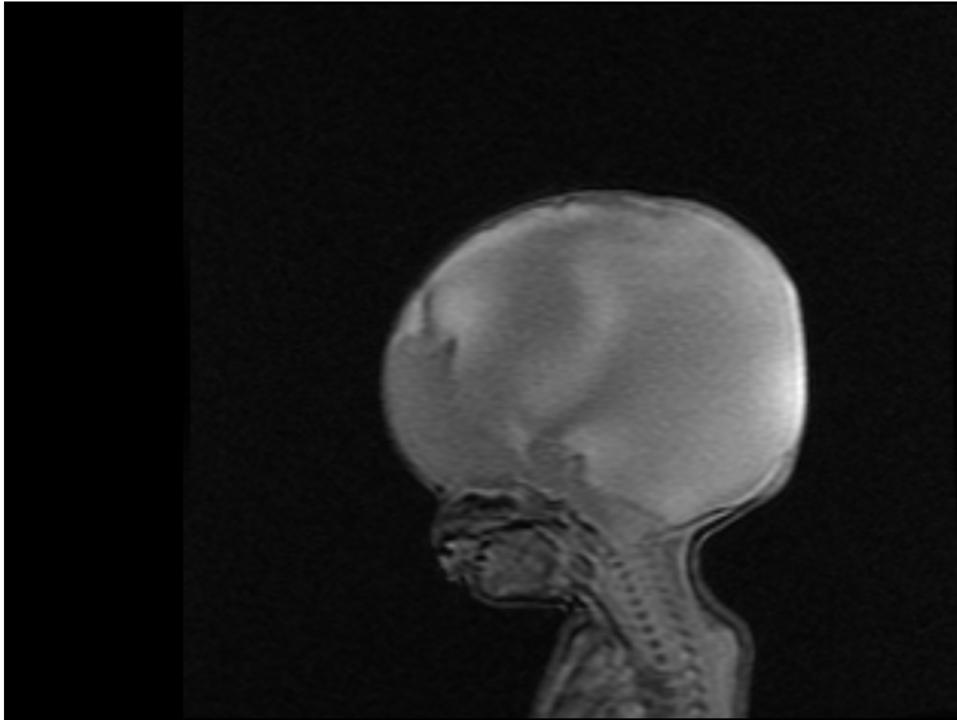
Admitted to NICU SVIA

CrUSS + exam confirmed AN findings

NGT feeds

MRI head: consistent with holoprosencephaly, probably of the semilobar or alobar type, discussed with neurosurgery

Case 2



Postnatal course

HC ↑ by day 7 - transferred to JRH

- D8 & D29: ventriculostomy with washout. Septo-optic dysplasia found
- D37: CONS meningitis
- D57: VP shunt inserted

Post operative complications: focal seizures, surgical-related DI, meningitis, suspected NEC

Transferred back to Wexham Day 65

Case 2

Current issues

Now 8 weeks post-term

SVIA
Feeding support – mainly NGT, SLT involvement
Gaining weight
HC stable, no seizures noted
Difficulties with positioning, physio involvement
Community nurse support for discharge

Concerns re parental visiting and capability of managing medical condition at home

Case 2

Communication challenges

Antenatal:
Early joint meeting
Plan for delivery
Life expectancy

At delivery:
Bedside / in theatre
1st discussion in NICU
Where to admit?

During admission:
Expected outcomes
ACPs
Parental involvement

Follow-up:
Ongoing MDT input
Palliative care
Open access to ward

What was done well?

Case 1:

- ACP discussed and written when appropriate
- Full MDT support, good community nurse input, frequent local consultant reviews

Case 2:

- Initial paeds input, robust plan communicated to all team & family.
- Early scan and neurosurgical input

What could be improved?

Case 1:

- More clearly documented discussions.
- Earlier/planned involvement of paediatric team
- ? Earlier MRI
- Parallel planning
- Managing multiple appointments with different teams, and expectations of parents

Case 2:

- Work in progress with getting parents involved in current needs and discharge plan