Doncaster and Bassetlaw Paediatric Department
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Welcome to the fourth issue of '**The PEN'** – the Paediatric Education Newsletter. This is a monthly educational bulletin of cases, clinical questions and learning points from recent teaching.

MCQ (Based on recent case at DRI)

A 10Kg, 1y old born to consanguineous patents presented 3 times over a 1 month period with high fever without focus. On each occasion he had a very high CRP >100 and marked neutrophilia (N 31). He was treated with IV cefotaxime for between 5-7 days on each occasion and his fever and CRP settled.

In his past medical history he had delayed separation of the umbilical cord and had recurrent episodes of umbilical flare requiring 4 courses of antibiotics.

What is the most likely diagnosis?

A. Leucocyte Adhesion Deficiency
B. Di George Syndrome
C. Chronic Granulomatous disease
D. HIV
E. SCID

Learning Points from this month's departmental teaching

Upper GI bleed – Iain Marshall

1. Significant upper GI bleeds can be difficult to differentiate – reliable indicators include Hb drop of 20, raised urea, pallor, and melaena.
2. Octreotide manages variceal bleeding by reducing portal pressures and is a very safe and effective drug.
3. Portal hypertension is often silent and commonly presents with splenomegaly or variceal bleeding.

A case of a newborn seizure on COU – Janani Devaraja and Neil Lawrence

1. Perform a hypoglycaemia screen on patients with blood glucose<3.3mmol/L (for neonates less than 72 hours of age, its blood glucose <2mmol/L on blood gas).
2. If there is cortisol deficiency, start cortisol at 10mg/m2/day in 4 divided doses.
3. Initiate cortisol treatment first prior to thyroxine if there is a combined deficiency to prevent development of adrenal fatigue.

Nephrotic Syndrome – Natasha Ponniah and Simon Hardman

1. Steroid resistance nephrotic syndrome is considered as patients who do not respond (or show remission) to 4 weeks of daily steroids
2. No difference in time to relapse between a standard course of prednisolone compared with extended (16 week with tapered dose) as per PREDNOS study.

Article Links

Leukocyte adhesion deficiency-I: A comprehensive review of all published cases.

Author(s): Almarza Novoa, Elena; Kasbekar, Sanchali; Thrasher, Adrian J; Kohn, Donald B; Sevilla, Julian; Nguyen, Tony; Schwartz, Jonathan D; Bueren, Juan **Source:** The journal of allergy and clinical immunology. In practice; 2018; vol. 6 (no. 4); p. 1418 The journal of allergy and clinical immunology. In practice

Dynamed: Part of Primary Disorders of Phagocyte Function
https://www.dynamed.com/condition/primary-disorders-of-phagocyte-function#MANAGEMENT_OF_LEUKOCYTE_ADHESION_DEFICIENCIES

Leukocyte Adhesion Deficiency StatPearls, Last Update: July 5, 2020.
https://www.ncbi.nlm.nih.gov/books/NBK539770/

Answer to MCQ = A

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Approach to defining Stillbirth- Umberto Piaggio (ST8)

The definition of "stillborn child" in England and Wales is contained in the Births and Deaths Registration Act 1953 section 41 as amended by the Stillbirth (Definition) Act 1992 section 1 and is as follows: "a child which has issued forth from its mother after the 24th week of pregnancy and which did not at any time breathe or show any other signs of life" (Previously the threshold had been 28 weeks) Similar definitions apply in Scotland and Northern Ireland.

Causes of stillbirth

Maternal causes:

- Maternal infection
- Diabetes, thyroid abnormalities, hypertension, SLE, cholestasis of the pregnancy, renal disease, sickle-cell disease as well as others

Foetal causes:

- Poor foetal growth or intrauterine foetal growth restriction (IUGR), (related to numerous maternal conditions described above)
- Multiple gestation, congenital anomalies, genetic abnormalities and infection

Placental causes:

- Abruption, PROM, vasa previa, chorioamnionitis, vascular malformations and umbilical cord accidents

Signs of stillbirth

Antepartum:

- Clear documentation of no foetal activity, decreased maternal weight, or fundal height.
- Auscultation alone is insuffient to confirm death, as well as easily misinterpreted (Boero Sign- hearing maternal aortic beat)
- Real-time USS is the gold standard for antepartum stillbirth diagnosis as there is a direct visualization of the absence of cardiac activity, absence of aortic activity and the absence of movements of the body or limbs of the foetus.

Postpartum:

- Apgar scores of 0 at 1 and 5 min
- Absence of signs of life* (**spontaneous movements, spontaneous respirations, and spontaneous cardiac activity**)
  - Heartbeats are to be distinguished from transient cardiac **contractions**
- Respirations are to be distinguished from transient fleeting respiratory efforts or gasps.
  - Appearance of the foetus may show signs of maceration and the level of maceration can determine time of death.

Article Links

The investigation of stillbirth, House of Commons Briefing Paper Number 08167 28 March 2019

Greentop Late Intrauterine Fetal Death and Stillbirth Green–top Guideline No. 55 October 2010

Tavares da Silva et al, "Stillbirth: Case definition and guidelines for data collection, analysis, and presentation of maternal immunization safety data", Vaccine. 2016 Dec 1; 34(49): 6057–6068.doi: 10.1016/j.vaccine.2016.03.044

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