Red Blood Cell Disorders Committee

BHS.be

MEETING October 26, 2017

7:00 PM

Partners present:

Laurence Dedeken, Phu-Quoc Lê, Olivier Ketelslegers, Marie-Françoise Dresse, , Pierre Philippet, Anne-Sophie Adam, Alina Ferster, Marie-Agnès Azerad, Philippe Maes, Lucie Pecheux, Ann Van Damme, Annelyse Bruwier, Cécile Boulanger, Veerle Labarque , Béatrice Gulbis

Guest: François Eyskens

Report:

- Updated recommendations: « Management of sickle cell disease » – Bram De Wilde

Discussion – suggestions :

Evaluation of transcutaneous saturation every 4-6h or continuously if respiratory symptoms or back pain

Fever : must be defined
> 38°C if < 2 y
> 38.5°C if < 4 y
> 39°C any age

AB : amoxycillin, always keep at hospital ; oral if very young children and depend on vein access ; not if clinically probably a viral infection ; otherwise all patients should be considered at high risk (see also no information available, no personal history or rapid degradation.
Splenic sequestration : no antibiotics
Liver sequestration : antibiotics (see origin not always known)

Pain : Be sure that for PCA the anesthetist doesn't give only a bolus
Add MEOPA (20') before morphine could be instaured
Auto and hetero evaluation are mandatory for a good evaluation of pain.
Be aware that in patients who experience a lot of pain episode, auto evaluation might be unreliable.

Hydratation : if drinking is possible, the better option ; if not, the survey of fluid balance is mandatory

Transfusion : No if simple VOC ; Ok for other events stated in the slideds + priapism
Exchange ++ mandatory if Hb level >9 g/dl, in case of CNS event or acute priapism.
Anemia and recognition of palor if black skin : palm of the hands ,soles of the feet, evaluation by the parents)
If Hb < 6 and Transfusion within the two last weeks : check for delayed hemolytic reaction.
ACS no transfusion if without any severe state (defined as Pa02 > 70 mmHg at ambient air)
CVA : keep the patient in your ward (not stroke clinic!!) ; HbS < 30%

Follow-up outside the hospital : work with « GP » paediatricians ; patient dependent, not easy to define.

- Clinical case
  5 y.o. HbSβ° (Syria)
  Hb ± 9.5 g/dL
  Transcranial doppler velocity very high 260-280 m/sec. It decreased wihout being on HU.

Such high velocities unlikely as relatively high Hb level. Check if velocities expressed as peak systolic velocities or TAMX. If abnormal TAMX → Treatment proposed : go to exchange transfusion at least 12 – 18 mois before bridge to HU alone (if velocities TAMX < 170 cm/ sec).

- Annual meeting on SCD – Lucie Pecheux
  The preliminary programme is presented and discussed
  Date of the meeting : 22/02/2017 – Location : CHU Saint Pierre Auditorium Bastenie

- Follow-up of EuroBloodNet - Béatrice Gulbis (see annex 2)

- Follow-up of the registry - Alice Ferster/ Sarah Wambacq (see Annex 3)

- Gaucher disease – protocol for screening – François Eyskens
  Proposal : to participate in a study on patients with splenomegaly with or without thrombopenia and without a diagnosis.

- « Emergency passport » – Novartis (see annex 4)
  - Opinion of the group :
    - Useful if people move from one hospital to another, to inform medical staff that perform the "triage".
    - To be given at the front desk (triage) but a link to a website that give access to the recommendations for care should be added on the passport
    - Add warning: "Don't overtransfuse"
    - Add that "No delay before taking the patient in charge"
    - In Antwerpen they will soon provide to the patient a USB stick containing all the information needed.
    - Partners suggest to develop an "App" for the patients.

Next meeting Thursday January 25, 2018.