How to assess and manage autoimmune haemolytic anaemia

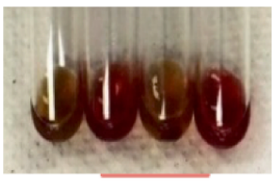
How do you spot children with haemolytic anaemia? Children with jaundice, pallor and passing cola – colour urine; may be shocked

Initial Investigations for AIHA: FBC, U+E, SBR (split), LFTs, reticulocytes, Group+DAT, film, LDH, haptoglobins, Infectious disease screen.

DAT+ve for C3: D-L test

What is the Donath-Landsteiner (D-L) test? Looks for the D-L (IgG) antibody that binds to P antigen of RBCs & fixes complement at low temperatures: then activates complement at core temperatures (37°C) causing red cell lysis .

Time taken for test: 24hr. Needs 2ml blood, kept at 37°C in thermoflask.: pre-warn the labs. Cost ~£108



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Test: After centrifuging patient's blood (37°C), Reagent P antigen positive group O RBC and donor serum (complement source) added. Sample split to 4tubes. Tube 1 at 37°C (90min) Tube 2: ice for 30min,then 37°C (60min). Tube3 (with 1%papain to expose more RBC P antigen) 37°C for 90min. Tube 4 (with 1%papain) : ice for 30min,then 37°C (60min).

Result : A positive D-L test ☑ occurs when haemolysis has occurred: tube 2&4 (ice then 37°C) and tubes 1&3 are negative ☒. This confirms PCH.

Real-world results:

Largest study (n=52 patients with PCH): 51 patients had positive D-L test, in 4 patients the significance of the positive test was unclear. Similar results from smaller case-series.

Clinical uncertainties

False negatives: when D-L antibody only at low levels.

False positives: Patient with cold agglutinins and IgM with I specificity (2% of patients with cold agglutinins)

Other issues: Identifies PCH but not the trigger.