Use of Rare Blood in Complex Cases

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Use of Rare Blood in Complex Cases

- The International Rare Donor Panel (IRDP)
- The role of the IBGRL Red Cell Reference department
- 5 complex case studies where rare blood provision was required



WHO International Rare Donor Panel (IRDP)

International Rare Donor Panel

- Rare Donor Panel was set up 47 years ago
- Has always been administered by the IBGRL Red Cell Reference department
- First edition was published in 1968 when there were 300 donors from 10 countries
- Currently we have 5264 donors listed, from 26 countries
- Not <u>all</u> donors listed independently

IRDP Collaboration



- UK National Frozen Blood Bank, Liverpool
- NBS South London
- Frozen Blood Banks, Amsterdam, Paris, Spain
- American RDP
- Japanese Red Cross RDP
- South African RDP

ISBT Rare Donor Working Party

International Rare Donor Panel

What do we do in Bristol?

- Compile and maintain information on rare donors from around the world that other centres have identified
- Keep up to date data on blood centres, donors and contact personnel
- Make information available to other blood centres via internet
- Co-ordinate requests when required



Case study EP

- Blood samples from patient (EP) referred from Sweden
- Third pregnancy
- Antibody reacting with all cells tested
- ?? Antibody specificity



Panel Cells	IAT Untreated	IAT Papain
1	3	4
2	3	4
3	3	4
4	3	4
5	3	4
6	3	4
7	3	4
8	3	4
9	3	4
10	3	4
Patient	0	0

Blood Provision for EP

- Higher incidence of Di^a in South American Indians (36%), therefore Di(b-) phenotype higher prevalence.
- Although referral from Sweden the name of the patient indicated the ethnic origin most likely South American
- Di(b-) units obtained from Brazil

Case study EJ

Panel Cells	IAT Untreated	IAT Papain
$R_1^w R_1$	0	0
2	4	4
3	4	4
4	4	4
5	4	4
6	4	4
7	4	4
8	4	4
9	4	4
10	4	4
Patient	0	0

- 73 year old English female patient for double knee replacement surgery
- R₁^wR₁ phenotype
- Serum strongly positive with all panel cells
- Anti-c+E+K+??
- Referred to IBGRL for identification of other antibody (ies)





Results

- <u>Patient's cells</u> and compatible panel cells were homozygous for low incidence C^w antigen
- Rare R₁^wR₁^w phenotype (Rh:-51)
- <u>Serum</u> contained anti-Rh51 +K
- Anti-Rh51 negative with :

 $R_1^{W}R_1^{W}$, -D-/-D-, Rh_{null}

Case study EJ

Rh antibody - presumed to be clinically significant

Possible options?

- R₁^wR₁^w,-D-/-D-, Rh_{null} very rare phenotypes
- One R₁^wR₁^w (K-) donor on UK National Rare Donor Panel
- ? availability of compatible panel cell donor
- Autologous (too old)
- Rh: -51 donors in Finland (i incidence of C^w)

Blood Provision for EJ

Actual transfusions

- One R₁^wR₁^w, K- unit obtained from NDP
- Compatible panel donor (SNBTS) called in to give a unit
- Both units transfused without incident
- Two units obtained from Finland for repair operation one month later following a fall

Case study VW

- 82 year old female patient (VW) with ischaemic heart disease, blood required urgently
- Blood samples referred to IBGRL red cell reference lab from South Africa
- VW had strong antibody reacting with all cells tested
- ? Antibody specificity



- Patient first tested in 1999
- Anti-Fy^a + unidentified antibody
- Transfused 3 units
- Sept 2011 transfused 3 units Fy(a-)
- Oct 2011 all cells incompatible

Case study VW

- Eluate of antibody [off Fy(a-) cells] was compatible ONLY with Rh_{null}
- D--/D-- cells incompatible
- Conclude: Rh–related antibody (+ anti-Fy^a)
- Rh phenotype : dce/dce [no D gene to sequence]
- Sequence *RHCE* gene

Case study VW

- Sequencing of her *RHCE* gene revealed a novel homozygous mutation in exon 1 (114A>C) giving rise to a Leu38Phe amino acid change
- This mutation is in the same region as mutations for C^w(Gln41Arg) and Cx (Ala36Thr)
- Homozygous C^w and C^x lack high incidence antigen Rh51

VW cells Rh:-51

RHD & RHCE exons 1-10





Transfusion Support for VW

- Referring laboratory had also found Rh_{null} cells to be compatible whilst we were working on this case!
- Received one unit from a local (South African) Rh_{null} [Fy(a-)] donor
- The patient required no further transfusions
- Complex investigation time consuming to solve

Case study Baby R

- 15 day old baby strong positive DAT, Hb 5.6g/dl, blood required urgently
- Blood samples from mother (African origin) referred to IBGRL from Lisbon,Portugal

Clue

- Mother had strong antibody reacting with all cells tested
- ?? Antibody specificity



Results

- Mothers serum was found to contain anti-C+hr^s (e-related 'high' - found in Blacks)
- Only blood compatible and suitable for baby was Rh_{null}, D--/D-- and Ro hr^s-
- Rare blood groups
- Mother (group B) and baby (group O)



Where are they?

- Only 3 group O Rh_{null} donors listed on International Rare Donor Panel (IDP)
- Few D--/D-- listed but >30 in Japan and also frozen units
- Some hr^s- in USA
- Invariably hr^S- supplied from South Africa

Case study Baby R

- New Rh_{null} donor recently recruited to the International Rare Donor Panel
- We contacted the Northern Ireland Blood Transfusion Service (NIBTS)
- NIBTS immediately contacted the donor and she went directly to donate a unit of blood
- Unit delivered in person by NIBTS staff member to Lisbon!



Baby was transfused with Rh_{null} blood

Hb 11.8g/dl 3 days post transfusion and baby continued to thrive

Case study Baby R

5 months post transfusion

- Baby had Hb of 11g/dl
- DAT negative
- Antibody screen negative
- Normal development



- There is only one group O Rh_{null} UK donor (KS)
- She gave the emergency unit for Baby R
- Gave a second unit specifically for another patient one year later
- Two units were frozen at the UK National frozen blood bank

- In April 2011 we were notified that KS was 24 weeks pregnant with a Hb 6.6dl
- Blood may be needed imminently and for delivery
- No frozen units available in UK
- We contacted several overseas centres for Rh_{null} availability

- Two donors were located and put on standby
- One from South Africa with Hb 12.2g/dl was put on medication to boost her Hb
- ? Bleed at slightly low Hb if necessary
- One from Brazil
- At 36 weeks gestation the situation became more urgent

- Patient had a large uterine fibroid and complications of delivery were anticipated
- The 2 units on standby were requested to cover surgery
- Both the South African and Brazilian donor units were shipped to N Ireland

Outcome

- Healthy baby delivered by CS at term
- No blood was required
- Rh_{null} units frozen at the National Frozen blood bank
- Effectively replaced the 2 units of KS that had been used for another patient!

Use of Rare Blood in Complex Cases



- Antibody ID in complex cases is often time consuming
- Blood provision is difficult and may be distant
- IRDP enables effective exchange of rare blood between countries
- Rare blood is required rarely!
- International cooperation ensures that blood is made available to specified patients



• The IBGRL Red Cell Reference team

- Everybody involved in Rare donor recruitment &rare blood provision
- The Donors!

The effort is appreciated

