Author's response to reviews

Title: A Neonatal Presentation Of Factor V Deficiency: A Case Report

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Version: 3 Date: 13 November 2006

Author’s response to reviews: see over
Dear Editor,

We have revised the manuscript following the recommendations of the reviewers and resubmitted a revised version. In the following a comprehensive reply, point by point, to the reviewers comments highlighting any changes made to the manuscript and giving reasons where recommendations were not followed.

Reviewer: Paula Bolton-Maggs:

General:
The English of the report has been improved and sentences shortened where required. A table summarizing the cases reported in the English literature so far has been compiled. We felt that it is beyond the scope of a case report to summarize all the other numerous rare coagulation disorders which would not do these disorders justice and would require a review article. We have mentioned that in factor XIII deficiency coagulation tests would be normal.

Specific comments:
1. We have highlighted in conclusion in abstract and in the main text the message that unusual bleeding in an infant should prompt coagulation studies particularly in consanguinity. The differential diagnosis of nipple bleeding was incorporated. We felt the picture of the bleeding nipple demonstrating the absence of features of differential diagnoses was important for documentation.
2. We have mentioned the general issue that unusual bleeding in an infant of consanguinous parents need to prompt coagulation studies.
3. We have deleted the sentence as requested.
4. The anticoagulant functions of factor V were not pertinent to our case and were therefore not mentioned.
5. We have replaced the word “heralded” by the word “preceded”.
6. We have mentioned method, normal range, result of sibling screening, and acknowledgement of genetics laboratory.
7. We have mentioned viral inactivation of FFP, haemostatic level of factor V, and maximum level of factor V achieved. We have explained the platelet transfusion and factor VIIa application. Both are empirical adjuncts used to optimise clotting in the intra-and postoperative period. Factor V after FFP application was checked and a maximum of 0.22 IU/ml just below the safe haemostatic levels achievable with 20ml/kg of FFP (20 min after application). We have mentioned the half life of factor V in the conclusion. We have mentioned that FFP was given every 48 hours and that factor V inhibitor assays were done whenever factor V levels were undetectable.
8. We have discussed the difficulties with adequate prophylaxis of bleeding manifestations in factor V deficiency. We discussed that prophylactic FFP application without evidence of severe bleeding manifestations cannot be justified at present and that genetic studies in the future may be able to identify infants at particular risk who could qualify for such a regime.

Reviewer Anil Pathare

1. We have shortened the abstract. Repetition of essential data is essential in the case presentation section of such an abstract.
2. We have omitted this term as recommended.
3. We have changed from “heralded” to “preceded”.

4. We have discussed the difficulties with adequate prophylaxis in these circumstances in the conclusions section and rephrased the sentence.

5. We have mentioned the half live of factor V as reason for the requirement of frequent FFP transfusions.

6. We agree and have deleted this paragraph as requested. We have raised the issue of uncertainty of clinical course.