



SDI Review Form 1.6

Journal Name:	International Blood Research & Reviews
Manuscript Number:	2015_IBRR_18559
Title of the Manuscript:	SEVERE HEMOLYTIC ANEMIA DUE TO de novo HEMOGLOBIN SABINE IN AN ARGENTINIAN NEWBORN. FIRST CASE IN SOUTH AMERICA
Type of the Article	Case Study

Gene

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PART 1: Review Comments

	Reviewer's comment	Author's comment (if agreed with reviewer, correct the manuscript and highlight that part in the manuscript. It is mandatory that authors should write his/her feedback here)
Compulsory REVISION comments	It is an interesting case report concerning the rare unstable haemolytic anemia. But discussion must be enriched more as which is the outcome of the patients, splenectomy has an impact in severe form, folic acid supplementation, what is happening in homozygous state? Correct the date of reference no 5.	Numerous cases of hemolytic anemia have been described that are a consequence of the presence in the erythrocyte of unstable hemoglobins. Most unstable hemoglobin variants in heterozygous state have a variable percentage of the variant. In our case it was 8% of total hemoglobin. This is a much smaller proportion than that found for most Hb variants, presumably because it is precipitated both <i>in vivo</i> and <i>in vitro</i> during manipulation in the experimental procedures. The erythrocyte containing Hb Sabine, have a markedly shortened survival as a consequence of impaired metabolism, probably due to dissociation and precipitation of abnormal beta polypeptide chains of Hb Sabine. Unstable Hbs are inherited in an autosomal dominant fashion, and virtually all affected individuals are heterozygous. The homozygous state for the unstable variants has not been found. Our patient, as all Hb Sabine carriers reported so far had significantly increased Hb F levels (HbF:10%) Patients with higher levels of HbF as those referred by Pavlovic S, et al. (8) and Panagoula Kollia, et al. (15) one had an association with the Xmn I polymorphism at -158 Gγ and the other a compound heterozygosity with non-deletional hereditary persistence of fetal hemoglobin respectively.



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		Almost all reported patients suffered from severe hemolytic anemia and were splenectomized at early age. Our patient that now is 17 months is been managed with folic acid and when she was 14 months received one transfusion. Splenectomy can not be predicted now, it will depend on the transfusion regimen, hypersplenism and how she passes her early childhood. In Pediatrics is ideal to splenectomize after the age of 6. Even though Hb Sabine is detectable by electrophoresis, its correct characterization requires more sophisticated methods, like high performance liquid chromatography, capillary electrophoresis or molecular analysis. The so far published cases of Hb Sabine in 1969 (4), 1983 (5), 1992 (6) and 2008 (12) were all de novo presentations. The only inherited case reported in 2004, was the son of a Yugoslavian patient (8).
<u>Minor</u> REVISION comments		
<u>Optional/General</u> comments		