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Acute splenic sequestration in children with sickle cell anemia

Dear Editor,

I am very pleased to congratulate the authors and the journal for the publication of the article "Acute splenic sequestration in a cohort of children with sickle cell anemia."[1] Since this topic is related to medical emergency, it is very important for all of us who work in general pediatrics. Other strengths of the study are the facts that it presents national data and, more importantly, what occurs in the context of the Brazilian public health services. I believe it is crucial to consider such aspect that has an overwhelming influence on good medical practice.

I also would like to emphasize the fact that, at emergency rooms, the diagnosis of acute splenic sequestration (ASS) is not always established based on the classic definition presented in the article – "sudden increase in the splenic volume, associated with a drop of at least 2 g/dL in hemoglobin concentration and reticulocytosis." Some of the factors contributing to that are: 1) relatives not always take to the emergency department the child's clinical and baseline hematologic data reports provided by hematology referral centers; 2) lack of computerized medical record available to all medical care units, what could solve the previously mentioned problem; and 3) the fact that sometimes it is not possible to perform reticulocyte count at many emergency department laboratories. With regard to the item 3 specifically, the results shown in Table 1 of the above mentioned article suggest that this was also a difficulty in the cases reported by Rezende et al.: hemoglobin concentration was measured in 80% (138/173) of the ASS cases, whereas reticulocyte count was performed only in 14% (24/173) of them.

In face of these limitations, the diagnosis of ASS is established based on clinical parameters such as significant splenomegaly, pallor and hemodynamic instability, in addition to very low hemoglobin levels.

The following consequences arise from that:

- Severe cases of ASS will not be underdiagnosed and will receive appropriate treatment at the emergency department. At discharge, patients will be sent to the referral center, in order to define the follow-up management.
- These extreme criteria are not sufficient to diagnose mild cases of ASS. Within the age group with the highest risk for ASS, splenomegaly, anemia and reticulocytosis are typical findings of underlying disease, even if there are no complications. If the child's clinical and baseline hematologic data are not available for the emergency department pediatrician, a mild case of ASS might be misdiagnosed as possible exacerbation of typical findings, triggered by infection, an event which these patients are more prone to. Even though it may not compromise the emergency care, in such situation, underdiagnosis certainly results in inappropriate follow-up of the patient, who will not receive the best care – attentive follow-up, periodic transfusion program or indication of splenectomy.
- There might be a diagnostic confusion between ASS and red cell aplasia if reticulocyte count is not performed at the emergency department before blood transfusion. This might cause under or overestimation of the actual frequency of ASS.

I would like to know if some data were analyzed in this population and not presented in the study:

- Were caregiver's educational level and family's town of residence (and access to the health care facility) associated with the case-fatality of ASS?
- Did pneumococcal vaccine protect patients against ASS?
- Was parvovirus B19 serology performed in any of these reported cases?
- And finally, a question not related to the main topic of the study: is the Neonatal Screening Program of Minas Gerais able to offer any kind of counseling to the relatives of children diagnosed with sickle cell trait regarding occasional reproductive risks for the person in adulthood?

I congratulate the authors for the published data, and it will certainly contribute to encourage debate and increase knowledge about this topic.

Reference

1. Rezende PV, Viana MB, Murao M, Chaves AC, Ribeiro AC. Acute splenic sequestration in a cohort of children with sickle cell anemia. J Pediatr (Rio J). 2009;85:163-9.

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Authors' reply

Dear Editor,

We are thankful for Dr Andréa Nogueira Araujo's relevant comments about our article,[1] which demonstrates that the publication of this article has reached one of its goals, that is, to promote debate about this topic among pediatricians.

In fact, episodes of acute splenic sequestration (ASS), mainly the mild cases, are often not diagnosed. One of the difficulties we faced during data collection was failure in recording data on medical records during intercurrent clinical events, which are usually treated at emergency departments. This fact may be explained by a combination of several factors. In some cases, the physician who saw the child during the acute event did not provide a complete report to the assistant physician at the Hematology Center of Belo Horizonte (HBH), either because not all the exams had been requested or because the local structure did not allow for the exams to be performed. Reticulocyte count, for instance, which is one of the diagnostic criteria for ASS, cannot be performed at night, on weekends and holidays at several emergency departments linked to the Public Brazilian National Health System (Sistema Único de Saúde, SUS). In other cases, the medical report was handed to the patient's relatives, who did not take it to the next follow-up medical visits at HBH. Finally, even when the report was taken to the medical visit, the assistant physician occasionally did not take note of all details about the acute episode on the medical record. To a certain degree, this lack of information limits data analysis in retrospective studies. In addition, the absence of such data may, as pointed out by Dr. Andréa, compromise the patients' appropriate clinical follow-up.

The variables "educational level of the child's caregiver" and "family's town of residence" were not analyzed regarding the case-fatality of ASS. Distribution of ASS cases by town of residence showed a predominance of patients from the countryside (77.5%) when compared with those from the state capital (22.5%), which was very similar to the distribution of the general sample studied (23.9% lived in Belo Horizonte and 76.1% lived in the countryside). However, with regard to the place where the medical care was provided for ASS episodes, we found a prevalence of medical care provided in Belo Horizonte in comparison with medical care provided in the countryside. This information exposes the weakness of the current health system decentralization, whose structure is often insufficient to provide adequate primary care to patients with sickle cell anemia.

In terms of antipneumococcal immunization, it was not possible to assess an occasional association with presence or severity of ASS. Currently, the vaccination calendar recommended in Brazil includes the basic scheme vaccines of the Ministry of Health and the complementary vaccines recommended by the Brazilian Society of Pediatrics. Therefore, there were prescription of antimicrobial prophylaxis and indication of special vaccination for all patients, according to the Protocol for Carriers of Sickle Cell Syndromes.[2] Nevertheless, we did not check with the families if the scheme was completed according to the assistant physicians' recommendation.

Serology for parvovirus B19 was not performed for the patients studied because this test is not available in our outpatient routine procedure.

With regard to the child with sickle cell trait, the Neonatal Screening Program of Minas Gerais (Programa Estadual de Triagem Neonatal de Minas Gerais, PETN-MG) has established a routine procedure of referring the newborns with sickle cell trait and his/her relatives to the referral primary health care unit, which is responsible for providing counseling related to the favorable clinical progress in these cases and to the occasional reproductive risks for the child in the future, as well as for providing genetic counseling to parents with respect to later pregnancies.

We also would like to highlight that a broader health policy is very important, with the focus not only on the treatment of the sickle cell disease complications but also on the prevention and early detection of risk situations, as well as on the maintenance of an organized structure of the SUS, being able to provide global care to patients, including all their biopsychosocial needs.

References

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