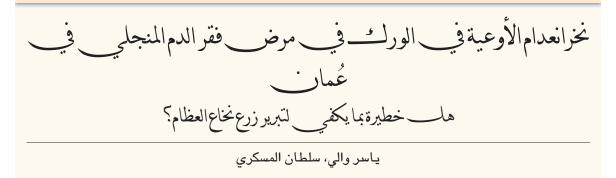
Avascular Necrosis of the Hip in Sickle Cell Disease in Oman

Is it serious enough to warrant bone marrow transplantation?

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Sickle cell haemoglobinopathy is the third most common genetic blood disorder in Oman.¹ The genetic background of the Omani population is heterogeneous and hence there are several haplotypes including African (e.g. Benin, Bantu) and the Arab-Indian resulting in different disease severity ranging from very mild to very severe.² In our population, there are some other factors that can modify the course of the disease like co-inheritance of alpha thalassemia that is highly prevalent in the Omani population (amounting to about 50%) and is believed to be associated with development of avascular necrosis (AVN) of the hip.³

AVN of the femoral head is a common complication in patients with sickle cell disease (SCD), and collapse of the femoral head occurs in 90% of the patients within five years of the diagnosis of osteonecrosis.⁴ In our hospital, we follow more than 500 children with SCD and 32 of them have developed AVN over the last 10 years. We have tried several treatment modalities, starting from early childhood, to improve or prevent progression of the disease in these patients. For milder grades of the disease, Steinberg stages I, II & IIIa (15 cases), we practice conservative measures such as non-weight bearing exercises using crutches, and physiotherapy including anti-gravity muscle strengthening exercises and a non-joint loading range of motion exercises.

For more severe forms of the disease (17 cases), many of our patients (11) have undergone surgical intervention either locally or abroad and came back to us for follow-up. Following published reports of some success, we used concentrated autologous bone marrow injection in 4 cases, with 3 failures and improvement of the hip in one child [Table 1]. In many of our patients (13/32), the disease has progressed to warrant hip replacement which is a major surgery. Since these patients are young, they may need to undergo revision 3 to 4 times in their life. We tried to address the reasons for the poor outcome in our children and we found that they do not use the crutches supplied and are not very compliant with the pre and postoperative physiotherapy rehabilitation programme.

Despite the seriousness of the problem and its life long effect on the patient's life with increasing pain, decreasing mobility, increasing liability to overweight, psychological stress and catastrophic economic impact on the patient, AVN in paediatric SCD patients has been inadequately addressed in the literature with only 19 papers cited in PubMed. In addition, there is no consensus in the literature on the best treatment options for the paediatric SCD patient with femoral head AVN. Though strong evidence is lacking, hydroxurea, despite

¹Department of Child Health, College of Medicine & Health Sciences, Sultan Qaboos University, Muscat, Oman; ²Department of Surgery, Sultan Qaboos University Hospital, Muscat, Oman. *Corresponding Author email: yasser_wali@hotmail.com its very positive effects on many aspects of SCD, has been implicated as a possible precipitator of AVN as it increases haemoglobin level in these patients.⁵ Nowadays, more indications are evolving for bone marrow transplantation (BMT) in SCD. However, the major problem for experts is to identify which patients require BMT earlier in life before the development of serious and life long complications.

The management of SCD–AVN, in our experience and in this part of the world,⁶ has been frustrating and associated in most instances with progression or recurrence of the disease. However, in Oman, we have many extended families with high rates of consanguinity thus increasing the possibility of human leukocyte antigen (HLA) matched sibling donors. The cost of an HLA matched transplant is much less in our setup (US \$45,000–50,000) than in Western countries.

Since SCD vasculopathy is the main factor for developing cerebrovascular accidents (CVA)⁷ and AVN,⁸ it is plausible to speculate that BMT will be useful in AVN as in the case of CVA. We believe that AVN of the hip is a severe complication of SCD that warrants haematology experts considering it among the indications for BMT.

Table 1: Outcome of different surgical interventions to treat sickle cell disease children with avascular necrosis of the hip in Oman

Intervention	No. of Patients	Result
Autologous bone marrow injection	4	1 success, 3 failures (progression)
Core decompression	4	1 success, 3 failures (progression to collapse)
Vascularised bone graft	2	1 success, 1 failure (non union)
Non-vascularised bone graft	1	Failure (progression to collapse)
Femoral osteotomy	1	Success
Pelvic osteotomy	1	Failure (progression to collapse)
Hip joint fusion	1	Failure (non-union)
Distraction arthrodiatasis	1	Failure (regional osteoporosis/collapse)

References

- 1. Al-Riyami A, Ebrahim GJ. Genetic blood disorders survey in the Sultanate of Oman. J Trop Pediatr 2003; 49:i1–20.
- 2. Al-Lamki Z, Wali YA, Wasifuddin MS, Zacharia M, Shakeel A, Rafique B. Natural history of sickle hemoglobinopathy in Omani children. Int J Pediatr Hematol Oncol 2000; 7:101–7.
- 3. Adekile AD, Gupta R, Yacoub F, Sinan T, Al-Bloushi M, Haider MZ. Avascular necrosis of the hip in children with sickle cell disease and high Hb F: Magnetic resonance imaging findings and influence of alpha-thalassemia trait. Acta Haematol 2001; 105:27–31.
- 4. Neumayr LD, Aguilar C, Earles AN, Jergesen HE, Haberkern CM, Kammen BF, et al. National Osteonecrosis Trial in Sickle Cell Anemia Study Group. Physical therapy alone compared with core decompression and physical therapy for femoral head osteonecrosis in sickle cell disease. Results of a multicenter study at a mean of three years after treatment. J Bone Joint Surg Am 2006; 88:2573–82.
- Sidani CA, Ballourah W, El Dassouki M, Muwakkit S, Dabbous I, Dahoui H. Venous sinus thrombosis leading to stroke in a patient with sickle cell disease on hydroxyurea and high hemoglobin levels: Treatment with thrombolysis. Am J Hematol 2008; 83:8–20.
- 6. Akinyoola AL, Adediran IA, Asaleye CM. Avascular necrosis of the femoral head in sickle cell disease in Nigeria: a retrospective study. Niger Postgrad Med J 2007; 14:217–20.
- 7. Hillery CA, Panepinto JA. Pathophysiology of stroke in sickle cell disease. Microcirculation 2004; 11:195–208.
- 8. Smith JA. Bone disorders in sickle cell disease. Hematol Oncol Clin North Am Dec 1996; 10:1345–56.