

Avascular Necrosis of the Hip in Sickle Cell Disease in Oman

Is it serious enough to warrant bone marrow transplantation?

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نخرانعدام الأوعية في الورك في مرض فقر الدم المنجلي في
عمان
هل خطيرة بما يكفي لتبرير زرع نخاع العظام؟

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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

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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 arthrodiastasis | 1 | Failure (regional osteoporosis/collapse) |

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