Newsbites

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babycell™
The Life Saving Cord Blood Bank

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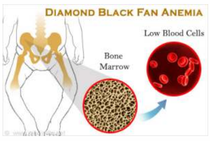
DIAMOND BLACKFAN ANAEMIA:

Diamond Blackfan Anaemia (DBA) is one of the rare group of genetic disorders, known as the Inherited Bone Marrow Failure Syndromes (IBMFS) [1]. DBA causes low red blood cell counts (anaemia), without substantially affecting the other blood components (the platelets and the white blood cells), which are usually normal.

Symptoms:

Approximately 40% of the patients are presented with one or more congenital defects. Most of these abnormalities are Cleft palate, high arched palate, thumb or upper limb abnormalities, cardiac defects, horseshoe kidneys, duplication of ureters, certain facial characteristics like tow-coloured hair (extremely blonde, almost white), a snub nose, wide-set eyes, and thick upper lip often appear in children with DBA. In addition, the head can be small with almond-shaped eyes and a pointed chin.



Treatment:

The cornerstones of treatment remain-

1.Corticosteroids (Medication):

The goal of corticosteroid therapy is to keep the haemoglobin at a healthy level. Doctors usually prescribe a certain dose of steroids for several weeks. Over time, the body might not respond to the corticosteroids and they will not work as well. If this happens, the doctor might increase the dose or suggest a different type of treatment such as blood transfusions or stem cell transplantation.

2.Chronic red blood cell transfusions:

Chronic blood transfusions might be recommended just as needed when the haemoglobin is lower than normal. Negative side effects of blood transfusion therapy can include blood transfusion reactions, infections, the development of red blood cell antibodies, and iron overload in different organs of the body.

HAEMATOPOIETIC STEM CELL THERAPY – The New Hope

Stem cell transplantation is the only definitive treatment for the haematologic manifestations of DBA[7]. Allogeneic matched sibling HSCT has been very successful8. For DBA patients, a Haematopoietic stem cell transplant is intended to restore the marrow's ability to make red blood cells.

Once the body starts producing red blood cells, the patient may experience a decrease in signs and symptoms of anaemia, such as tiredness and paleness. Often times, stem cell transplant may result in a cure of DBA and, when successful, may often extend and improve the quality of life of the patient. The person will no longer require long-term steroid medicine or blood transfusions.

Stem cell transplantation (SCT) has been explored as an alternative to chronic transfusions since 1976.[2] More than 70 DBA patients have undergone SCT so far[3,4,5] with an overall survival of about 85% at three years from sibling donors in more recent reports.[3,5]

CASE STUDY:

A nine-year-old boy of north Indian descent, the third of four siblings born to healthy, non-consanguineous parents was detected with DBA at the age of 8 months. Though initially steroid-responsive, the boy later became unresponsive to both steroids and cyclosporine. By the fourth year of life, he was completely transfusion-dependent, and iron Chelation was started only in the sixth year of life. He developed lower-limb weakness and calf muscle pseudo hypertrophy. The weakness was progressive in nature, and by the age of five years the boy required support to climb stairs. The child had an unaffected elder brother who was a HLA-identical match (6/6 antigen match) and underwent allo-HSCT treatment. This patient was evaluated by a neurologist pre-transplant and periodically post-transplant. The patient was wheelchair-bound pre-transplant, and, 45 months post-transplant, there has been no clinical deterioration whatsoever in the boy's motor power[6].

Sources:

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3.Willig T-N, Niemeyer CM, Leblanc T, TiemannC, Robert A, Budde J, et al. Identification of new prognosis factors from the clinical and epidemiologic analysis of a registry of 229 Diamond-Blackfan anaemia patients. Pediatr Res 1999;46:553-61.

4.Alter BP. Bone marrow transplant in Diamond-Blackfan anaemia. Bone Marrow Transplant 1998;21:965-6.

5.Vlachos A, Federman N, Reyes-Haley C,Abramson J, Lipton JM. Haematopoietic stem cell transplantation for Diamond-Blackfan anaemia: a report from the Diamond-Blackfan anaemia registry. Bone Marrow Transplant 2001;27:381-6.

6.http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3123644/

7.August CS, King E, Githens JH, et al. Establishment of erythropoiesis following bone marrow transplantation in a patient with congenital hypoplastic anemia (Diamond-Blackfan syndrome). Blood 1976;48(4):491-498.

8.Vlachos A, Federman N, Reyes-Haley C, Abramson J, Lipton JM. Haematopoietic stem cell transplantation for Diamond Blackfan anemia: a report from the Diamond Blackfan Anemia Registry. Bone Marrow Transplant 2001;27(4):381-38

International Make a Wish Day

Dear Mommies-to-be,

On the occasion of International Make a Wish Day (29th April), Babycell will not only secure your baby's stem cells but also fulfill one of your wishes, this whole month!

Choose your option

1. Photo frame + Mummy & Tummy Book
2. Baby Record Book + Medela Micro-Steam Bags with pouch
3. Baby on board stickers + Mom & Me Goody Bag

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EVENTS

Mummy & Tummy Sessions and Babycell's Mother Care Forum celebrates International Women's day with would-be mother's in various cities like Hyderabad, Pune, Mumbai.

Pune

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MCF Hyderabad

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