EXERCISE AND SPORTS IN HEMOGLOBIN DISORDERS

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- Inherited hemoglobin disorders ; the structural hemoglobin variants and the thalassemias , all are caused by defective globin production.
- Those with a single defective globin gene are carriers or heterozygotes, are symptomless.
- Those homozygous or compound heterozygotes for the disorder develop symptoms of the disease , mainly anemia.

- Evidence based guidelines developed for proper management of thalassemic patients , and the development of new oral iron chelators , all increased expectancy of a long survival of good quality for thalassemic patients.
- This encourages the patients to plan for their future, having a job, family, children , and other activities like exercise and sports.

- Lower hemoglobin content of red blood cells (RBC) and lower RBC production is responsible for inadequate oxygen transport to tissues.
- Severe anemia interferes with normal physical development, resulting in growth retardation.
- The management mainly takes the goal of adequate RBC transfusions (pretransfusional hemoglobin level kept around 10 gr/dL), allowing normal activity and quality of life.

- Insufficiently treated anemia will cause osteoporosis, insufficient muscle building, heart failure, frequent pulmonary infections, reduced exercise tolerance which are all necessary elements for a normal exercise.

Goals of transfusion

- Correction of anemia (sufficient to eliminate hypoxia)
- Suppression of erythropoiesis
- İnhibition of gastrointestinal iron absorption

- Sufficiently treated anemia, will cause iron accumulation in tissues, which when untreated will lead to heart failure and death, accumulation in endocrine organs will cause endocrine abnormalities which again interferes with growth, muscle and bone metabolism, and other endocrine abnormalities (diabetes etc.)

- Deferoxamine (DFO) toxicity: at high doses or at therapeutic doses in patients with hypersensitivity, can be toxic to osteogenesis, collagen synthesis, bone turnover, leading to reduced growth (especially trunk), sliding femoral head, valgus deformity, rachitic like radiological lesions.

- Bone disease in thalassemia is multifactorial (anemia, hypogonadism, diabetes, cardiac dysfunction, chronic hepatitis,deferoxamine toxicity,etc), and is already present in childhood. Prevention is the basis of management.
- Stopping smoking, a calcium rich diet, correction of hypogonadism, and exercising is recommended.

- Brittle bones will increase the risk of fracture during many types of exercising but especially with heavy weight lifting and resistance training.
- Exercising on a regular basis is an important point for maintaining healthy growth and development. İt improves oxygenation and muscle development if enough hemoglobin is present.
- For strong bones, brisk walking, jogging, running, aerobics, step, dancing, circuit training, weight bearing exercises will be suggested according to the person's medical condition.

- If the person is anemic, enough oxygen will not be supplied to muscles during exercise, resulting in tiredness and muscle pains.
- Thalassemic patients may be suggested to receive transfusions 2 days before known physical activity programs.
- Heart problems and restrictive type of lung problems are main hinderances against exercise capacity and mostly occur due to iron overload.

- Studies regarding lung function are contradictory.
- Cooper et al.demonstrated a restrictive pattern in some patients that tended to correlate with age- duration of the disease.
- Keens et al.described abnormalities in pulmonary function tests suggesting small airways diysfunction which could be due to iron deposits in peripheral airways.
- Several authors suggested the possibility of pulmonary vascular involvement by microthrombi.
- Then most studies have revealed either an obstructive but most frequently a restrictive pattern of lung pathology.

- C.Cracowski et al. (1998) investigated lung function, exercise testing and echocardiography a few days after transfusion and found a significant impairment in exercise capacity in adult thalassemic patients which was not observed in children. No evidence was found in favour of pulmonary vascular or parenchymal involvement responsible for exercise limitation. The abnormal cardiac response was suggested to contribute to the exercise limitation observed.

- There are some reports that immediately after transfusion, for thalassemic patients with a history of airway obstruction, had a worsening of their airway obstruction after transfusion. This was postulated to airway narrowing due to airway wall vascular engorgement or oedema induced acutely and aggravated after transfusion. (Santamaria et al. 1994, Villa et al.1996)
- Many studies didn't find a correlation between respiratory function impairment and body iron content (Dimopoulou et al. 1999, AM Li et al.2002), while some studies did (JM Factor et al.1994, Kanj et al.2000)

- Infiltration by abnormal erythroid cells (ineffective erythropoiesis) has been proposed to explain the decrease of the gas transfer factor of the pulmonary membrane (alveolocapillary membrane block) and decrease in total lung capacity and exercise desaturation (Aessopos A et al.1995), together with pulmonary artery disease; pulmonary hypertension (Grisaru et al.1990).

- There are cognitive and emotional factors leading behaviour for participation in the social life and exercise, as studied by many scientists in Italy, United Kingdom, Greece and India.
- Khurana et al.’s (2006) study showed adverse impact of of thalassemia perceived in sports in about 70% of study population.

CONCLUSION

- Today's management protocols have given us increased expectancy of a long survival of good quality for thalassemic patients.
- Bearing this in mind, thalassemic patients should pay attention to their medical treatments and always participate in social activities and sports. They should take care to maintain healthy life styles, with regular exercising, no smoking and diets sufficient for calcium, calorie, and protein.
- We wish healthy days for all of them.