Original Article

RESULTS IN OSTEOARTICULAR SURGERY OF THE HAEMOPHILIAC PATIENTS

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Abstract

Hemophilia is a hereditary disease due to a defect of chromosome X, which lead to impaired production of coagulation factor VIII in hemophilia A (85% of cases) and factor IX in hemophilia B.

The evolution of hemophilic arthropathy is almost always from haemarthrosis to chronic synovitis and extensive erosion of the articular surface and, ultimately, the final stage of joint destruction - chronic haemophilic arthropathy.

The present paper aims to analyse results obtained in treating 105 patients that received surgical care (a total of 107 surgical interventions) within the Department of osteoarticular surgery for haemophiliacs, of the Orthopaedics-Traumatology Clinic II Timisoara, between year 2001 and 2012.

The treatment protocol was performed in collaboration with the team of hematology specialists from the Clinic of Pediatric OncoHematology - Pediatrics Hospital"Louis Ţurcanu" from TimiŞoara.

With substitution treatment and correct surgical indication, osteoarticular surgery applied to hemophiliacs, in a specialized centre, by a multidisciplinary team, can lead to good results with acceptable risks.

Keywords: *haemophilia, chronic arthropathy, bleeding, rare diseases*

Rezumat

Hemofilia este o boala ereditara datorata unui defect al cromozomului X, care determina o productie deficitara a factorului de coagulare VIII in hemofilia A (85% din cazuri) si a factorului IX in hemofilia B.

Evoluţia artropatiei hemofilice este aproape întotdeauna de la hemartroză la sinovită cronică şi eroziuni extinse ale suprafeţei articulare şi, în cele din urmă, la stadiul final de distrugere articulară – artropatia cronică hemofilică.

Lucrarea de fata analizeaza rezultatele tratamentului aplicat pe un lot de 105 pacienți operați (în total 107 intervenții chirurgicale) în Compartiment de chirurgie osteo-articulară al bolnavilor hemofilici din Clinica II Ortopedie – Traumatologie în intervalul 2001- 2012.

Tratamentul chirurgical al hemofilicilor se realizează în colaborare în echipa cu Clinica de OncoHematologie Infantilă de la Spitalul de Pediatrie „Louis Țurcanu" din TimiŞoara.

Cu un tratament substitutiv Şi o indicație chirurgicală corecte, chirurgia osteoarticulară la hemofilici, efectuată într-un centru specializat, de către o echipă pluridisciplinară , poate da rezultate bune cu riscuri acceptabile.

Cuvinte cheie: *hemofilie, artropatie cronică, hemoragie, boli rare*

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INTRODUCTION

Hemophilia is a hereditary disease due to a defect of chromosome X, which lead to impaired production of coagulation factor VIII in hemophilia A (85% of cases) and factor IX in hemophilia B. There are described three types regarding the severity of the disease: mild, with a concentration of deficient factor greater than 5% of normal values, an average of a concentration between 1 and 5% and a severe, with more than half of cases of hemophilia, with a concentration of factor VIII or IX less than 1% of normal. Hemorrhages in muscles and joints occur spontaneously in severe form, running a minor injury in the moderate and only after a major injury or surgery in mild forms. (1)

The evolution of hemophilic arthropathy is almost always from haemarthrosis to chronic synovitis and extensive erosion of the articular surface and, ultimately, the final stage of joint destruction - chronic haemophilic arthropathy. End stage of arthropathy is complicated by severe limitation of joint range of motion secondary to the arthrofibrosis due to the replacement of the hypertrophic synovium with a dense fibrosis. Severe contractures, angular