



Introduction and Objectives

Hemophilia is a rare occurrence in females. Symptomatic hemophilia can affect females by one of the following mechanisms: (1) Extreme lyonization of the unaffected X chromosome, (2) hemizyosity of X chromosome in females with Turner syndrome (X0 karyotype), (3) homozygosity in female progeny of hemophilia carriers and affected hemophilic males. The Israel national hemophilia center treats 655 patients. 5 are females.

We hereby present a 28 years old female patient with severe hemophilia A undergoing full pregnancy and labor.

Materials and Methods

Our patient was diagnosed with severe hemophilia A at the age of ten months due to repeated hemarthrosis. She was later found to be a carrier of intron 22 FVIII inversion with extreme lyonization of the other X chromosome. Since diagnosis, she was treated with pdFVIII on demand. Once married, the options regarding conception and prenatal diagnosis were discussed with her and her husband. Yet, she became spontaneously pregnant, refusing any intervention, even after the fetus was found to be male.

Results

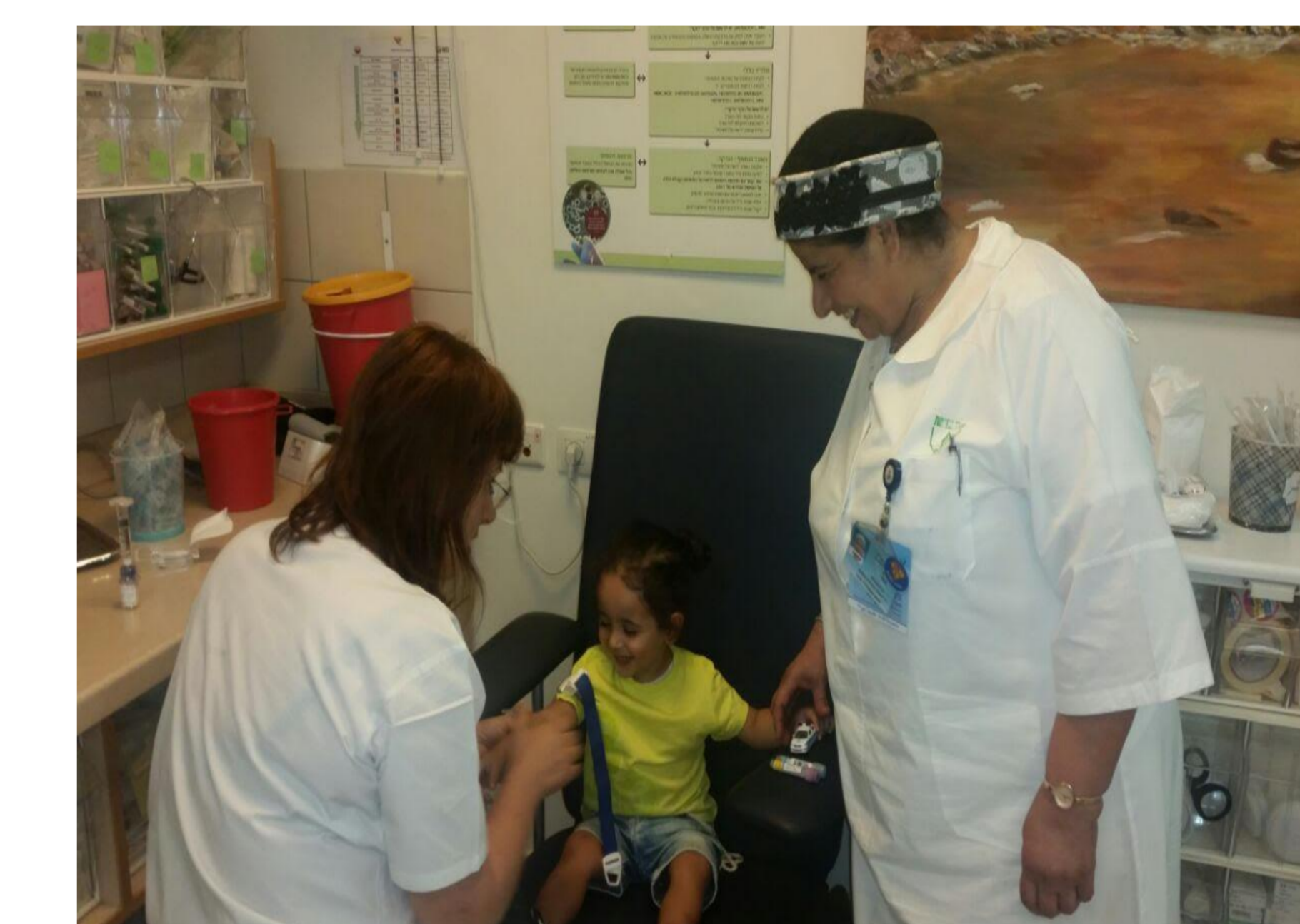
From early stage of pregnancy the patient received prophylactic FVIII, and was regularly seen both at our hemophilia center and at the high risk pregnancy unit. Towards labor, vaginal delivery was planned, with intent to go for caesarean section early in the course of any complication, avoiding instrumental delivery. However, due to pain intolerance, a semi elective caesarean section was performed under general anesthesia at 38 weeks gestation with application of factor VIII, tranexamic acid and fibrin sealants. FVIII activity was maintained at 80-100% on days 1-3 post delivery, 50% on days 4-7 and thereafter, regular prophylaxis was resumed. A healthy baby was born with severe hemophilia A. Both mother and child continue prophylaxis and both are doing well.

Conclusions

Pregnancy and labor in female patients with severe hemophilia A may be safely managed using a multidisciplinary team approach.



Israeli National Hemophilia Center entrance and kids reception area



Treatment of patient and child with factor VIII product at the center

* Pictures presented with the patients' approval