

The Lioness and the young lion – a case report

G. Goldstein, T. Brutman Barazani, D. Bashari and G. Kenet

The Israeli National Hemophilia Center, Sheba Medical Center, Tel Hashomer, Sackler medical school, Tel Aviv University, Israel

Introduction and Objectives

The Israeli national comprehensive care hemophilia center treats 655 patients (650 male and 5 females). Each of the female hemophilia patients has her own story. We decided to tell the story of one of them and her fight against all odds to become a mother

Materials and Methods

L.V is a 28 year old severe hemophilia female patient that was diagnosed at the age of 10 months with FVIII intron 22 inversion and extreme lyonization. As her social worker and her psychotherapist I was in contact with the family, kindergarden and school teacher and her parents. I was there beside her in good times and bad times, seeing her growing up being a teenager and then a young woman, later getting married and divorced during her pregnancy and delivery Table 1 presents the special unique challenges of women with hemophilia, as opposed to males with hemophilia or women who are hemophilia carriers. All those issues need to be addressed and require constant care and support of the psycho- social therapist.

Results

Table 1 presents the special unique challenges of women with hemophilia, as opposed to males with hemophilia or women who are hemophilia carriers. All those issues need to be addressed and require constant care and support of the psycho-social therapist.

LV attended a regular school and led active social life. Being a popular girl she danced in Ballet and Jazz companies. When she started dating boys she has been rejected many times due to Hemophilia and problems around her ability to deliver healthy children. She finally got married at the age of 24 years, divorced 2 years later (due to inability to have a child) yet got pregnant *(from her ex husband) soon afterwards and underwent full pregnancy and delivery as a single parent with my psychosocial support and under medial intensive supervision as high risk pregnancy case. 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. A baby was born with severe hemophilia A. Both mother and child continue prophylaxis at our center and both are doing well. The boy is well developed and very sociable.

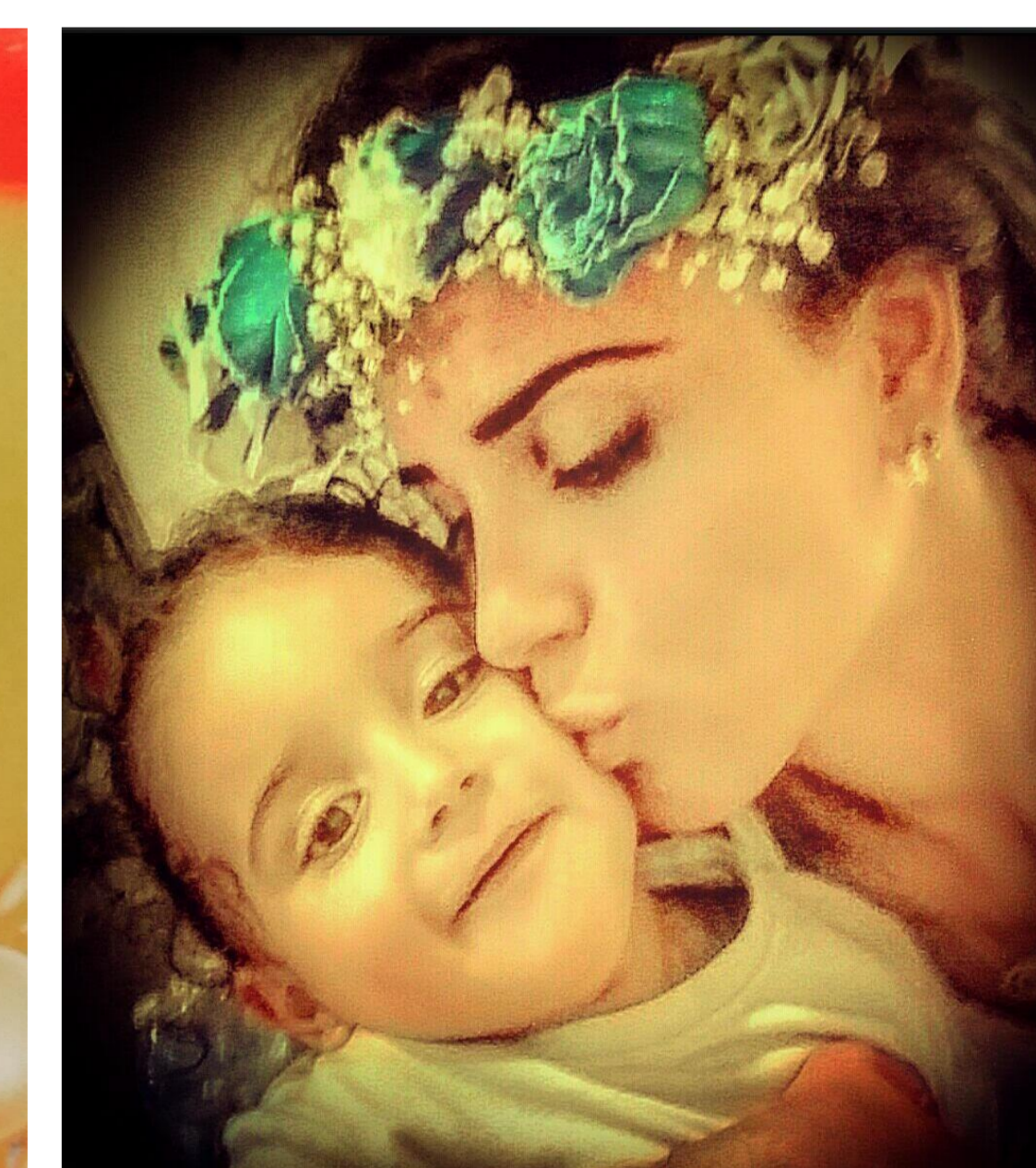
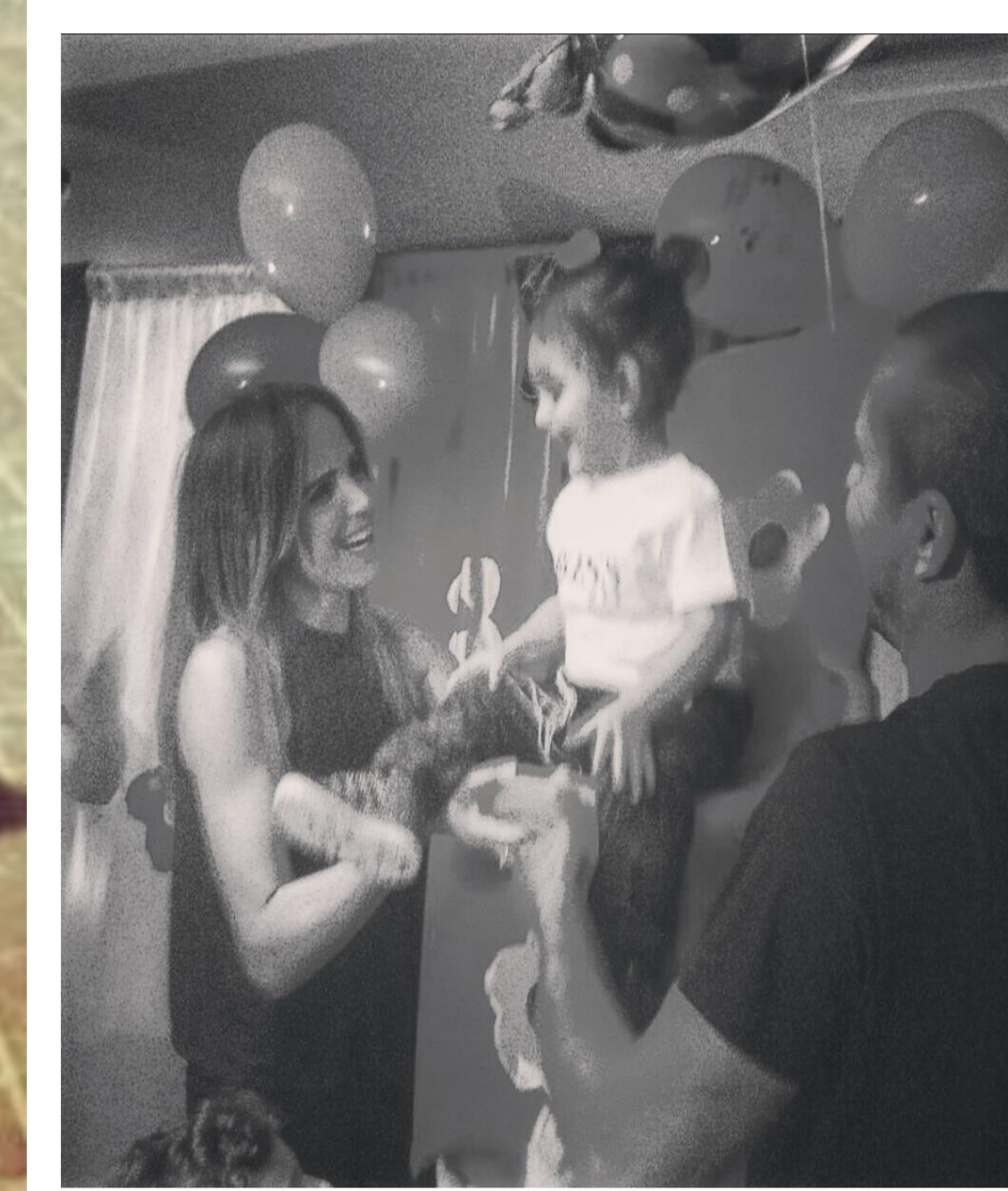
| | Male | Female | Carrier |
|-----------------|------------|--|---|
| Joint Bleeding | + | + | Usually no |
| Menstrual bleed | NO | + | + |
| Marriage | No problem | Problematic | No problem |
| Children | Healthy | May deliver boy with Hemophilia prenatal diagnosis is not optional | May deliver boy with Hemophilia, prenatal diagnosis is optional |
| Pregnancy | NA | Problematic | No Problem |
| IVF/ PGD | NA | Problematic | No Problem |
| Delivery | NA | May bleed | May bleed |

Table 1: Characteristics of women with Hemophilia as opposed to males or carriers of Hemophilia

Conclusions

LV is a unique rare case of a women with severe hemophilia A undergoing struggles of life since her diagnosis. While medical care has always been important, the administartion of Psychosocial support was crucial for her development and for success of therapy.

While receiving psychosocial support she underwent childhood , puberty, adulthood, marriage, divorce, preganacy delivery and parenting of a child with severe Hemophilia Now LV lioness with extreme lyonization is on a new battle for quality of life for her son and herself. She is now in the process of opening a business and building her independent future career.



Patient's approval was given to present the pictures

