

Experience of Self-infusion and Prophylaxis in Pediatric Hemophilia Patients

Introduction and Objective: Severe hemophilia is associated with bleeding into joints and development of arthropathy. Bleeding into a joint can cause pain, immobilization and irreversible arthropathy. The illness can prevent the patient from living an active and normal life. Prophylactic treatment with infusion of clotting factors to prevent chronic arthropathy is recommended for all patients with severe hemophilia. Evidence from studies demonstrates superior outcomes in prophylaxis compared to episodic treatment. In order to evaluate the practice of Prophylactic factor therapy and learning experience of self-infusion in pediatric patients with hemophilia, we performed a retrospective study from a single-center.

Materials and methods: A total of 34 severe hemophilia patients on prophylactic factor replacement therapy and their parents were analyzed. All data involving pediatric hemophilia patients on prophylactic treatment between 2014 and 2015 in a research hospital at Istanbul, were extracted from patient files.

Results: All patients had severe hemophilia A and started prophylaxis at a median age of 9.2 years. A majority (%94) of the children (32/34) started prophylaxis after a joint bleeds, and only two patients (%6) started primary prophylaxis. None of the patients developed an inhibitor on prophylaxis. Two of the patients who had inhibitors before prophylaxis, used Feiba (activated prothrombin complex concentrate) for prophylactic treatment. 12 of 34 (%35) patients accepted to have training for self-infusion. 9 of 12 parents of patients (%75), and one patient himself, successfully learned intravenous infusion. Overall, parents or patients needed a median of six visits to learn home treatment. In 83% of cases (10/12), the mother was the first who started learning to infuse the child. Four of 34 patient's parents (%12) refused self-infusion training and preferred to continue receiving prophylactic factor infusion at the healthcare center or hospital. In 16 case (% 47), himself or parents of patients was able to do self-infusion and didn't need any training also.

Conclusions: Early recognizing bleeding symptoms and self-care training help to achieve a high quality of life. Learning self-infusion of clotting factors requires time and effort. The difficulties associated with maintaining a prophylactic infusion regimen may result in patient nonadherence. Home treatment allows immediate access to clotting factor, hence optimal early treatment and must be supervised closely by the care team and should be initiated after adequate training. Early recognizing bleeding symptoms and self-care training help to achieve a high quality of life.

