A4. HEMOPHILIA MANAGEMENT – STILL A CHALLENGE IN CLINICAL PRACTICE?

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Hemophilia requires a life-long commitment to the regimen of care. In persons and families affected by hemophilia, adherence is more difficult when a young child must be treated every other day or 3 times per week during prophylaxis.

The primary difficulty with adherence to a regimen of prophylaxis in young children is venous access which is an essential aspect of hemophilia care. According with WFH Guidelines, veins must be treated with care because they are the lifelines for a person with hemophilia.

Peripheral venous access is strongly encouraged in the Consensus Recommendations, if veins are adequate and if the patient is old enough. Most people using peripheral venous access use butterfly needles. With this method, a single needle is used for each treatment and then removed. This means that nothing remains under the skin — which results in reduced risk of infection and increased freedom.

Venous access devices should be avoided whenever possible but may be required in some children. Infection is the primary complication associated with central venous access devices (CVAD) and the most common reason for their removal. The blood clots are also a complication of CVAD. CVAD can be complex and require significant training. There is also risk of the device breaking or malfunctioning. For all of these reasons, the Consensus Recommendations suggest that CVADs be used only when peripheral access is not feasible and only for as long as medically necessary.

Drugs that affect platelet function, particularly acetylsalicylic acid and non-steroidal anti-inflammatory drugs should be avoided.

Physical activity should be encouraged to promote physical fitness and normal neuromuscular development, with attention paid to muscle strengthening.

In conclusion, peripheral venous access should be preferred for administration of replacement therapy in hemophilia patients and patient and family education is also an important in order to increase patient adherence to treatment.