Medical Care for Athletes With Hemophilia

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Hemophilia is a hereditary disease characterized by impaired coagulability of the blood resulting from either decreased production or decreased functional activity of a coagulation factor. The most common hereditary coagulation disorder is hemophilia A, which occurs in about 1 in every 10,000 males. This disorder results from a gene defect on the X chromosome that causes partial or complete deficiency of Factor VIII coagulant activity. Because hemophilia A is an X-linked recessive disorder, it affects males, who only have one X chromosome, and is carried by females, who sometimes show a lesser tendency to bleed, because females have two X chromosomes and the disorder is recessive. Although hemophilia A is known as an inherited disorder, nearly 30% of individuals with it have no prior family history, in which case it is most likely the result of spontaneous genetic mutation. Hemophilia B (Christmas disease) and hemophilia C (Rosenthal syndrome) involve partial or complete deficiency of Factors IX and XI, respectively. These disorders are not as common as hemophilia A, occurring in 1 in every 100,000 males. Clinical symptoms for both conditions include hemorrhage and hemarthrosis.

Management Plan for Athletes With Hemophilia

Athletes with hemophilia are participating in competitive sports. The sports-medicine staff must be proactive in developing a management plan for their safe participation. The plan should include referral to a hematologist for preparticipation physical clearance and after every injury. Other components of the plan include desmopressin (DDAVP) effectiveness testing, availability of Factor VIII at all away contests, and knowledge of locations of hemophilia treatment centers near away-competition sites (see the sidebar).

Medical Clearance

Preparticipation Physical Examination

Past history of injuries, severity level, type of sport, and response to treatments all need to be taken into account before a final decision is made regarding participation status. Physical and emotional maturity should also be considered. Chronological age, Tanner stage, and muscle strength can be used to assess physical maturity. An individual who is aware of his or her body and does not take unnecessary risks is more likely to be successful in sports, whereas risk takers are more likely to sustain injury. The decision to allow participation should be made with the aid of the patient’s hematologist. The two primary considerations in determining the athlete’s participation status are the severity level of hemophilia and type of sport in which the athlete wishes to participate.
Severity of Hemophilia. There are three categories of classification of hemophilia based on the severity of the disorder. Severe hemophiliacs have less than 1% of the normal Factor VIII. These individuals frequently bleed from minimal or unrecognizable trauma, especially into joints and muscles. Moderate hemophiliacs have 1–5% of the normal Factor VIII concentration level and will hemorrhage from moderate trauma. Finally, mild hemophiliacs have 5–50% of the normal concentration level of Factor VIII. These individuals are at minimal risk for spontaneous hemorrhages but are likely to bleed after trauma or surgery.1,6,8

Type of Sport. There are three general categories of sport classification: collision, contact, and noncontact. In noncontact sports, contact between athletes is highly unlikely. Track and field and tennis are examples of noncontact sports. Contact sports have the possibility, but not the intent, of contacting opposing athletes. Examples of these sports would include basketball and soccer. Collision sports are played with the intent to strike opposing athletes. Sports in this category include football, hockey, and rugby.9 Sports that are associated with a higher degree of inherent risk of musculoskeletal injury might not be appropriate for a hemophiliac athlete.

The results of one study showed two major trends in team physicians’ opinions on hemophiliac athletic participation. As the risk of injury with each type of sport (contact vs. noncontact) increases, the willingness of team physicians to allow athletic participation decreases. In addition, as the severity of the disorder increases, the willingness of team physicians to allow athletic participation decreases. The results of this study indicate that the severity of hemophilia and type of sport dramatically influence the decisions of team physicians.9

Acute-Injury Management

The first recombinant FVIII (rFVIII) was derived in 1992. The genetically engineered Factor VIII allows for hemorrhage control with minimal risk of disease transmission. Because the rFVIII is formulated in human albumin, however, there is still some potential for disease transmission. A second generation of rFVIII that does not require the use of human albumin was produced in 1997.10 The creation of rFVIII has made treatment more readily available for hemophiliacs, and the current form allows for treatment with no risk of disease transmission.

Individuals with hemophilia A have been allowed to become more active with advancement in treatment for hemophilia. rFVIII enables an athlete to prevent or treat bleeding that occurs as a result of injury, giving the athlete reassurance that the injury can be managed efficiently. Plasma-derived concentrations are also currently used and carry a low risk of transmitting blood-borne infections. Methods such as selecting low-risk plasma donors, adopting polymerase-chain-reaction-based virus detection, and retesting donors 6 months after taking blood to determine whether it can be used have been employed to reduce the risk of disease transmission.10 Factor IX is also available in virally attenuated concentrations that can be administered to an injured athlete with hemophilia B.

Return-to-Play Guidelines

The return-to-play guidelines follow the same steps as for the other athletes, with a few modifications.

Management Plan for Athletes With Hemophilia

Medical Clearance
- Preparticipation physical examination
- Acute-injury management
- Development of return-to-play guidelines

DDAVP-Effectiveness Testing

Education of Sports-Medicine Staff
- General knowledge of hemophilia and injury management
- Individual management plan

Home-Contest Considerations
- On-site DDAVP (if effective)
- On-site Factor VIII

Away-Contest Considerations
- Location, directions, and contact numbers for nearest hemophilia treatment centers
- Transportation of DDAVP (if effective)
- Transportation of Factor VIII
An athlete with hemophilia who experiences irregular hemorrhages or hemarthrosis secondary to injury must be managed conservatively. Rehabilitation of an athlete with hemophilia would follow the same theories and guidelines as for other athletes. The primary concern is to gauge the inflammatory response to ensure that the therapeutic exercises do not interfere with the healing process. Clearance for any injury that involved this type of bleeding must be initiated by the team physician in collaboration with the hematologist and the certified athletic trainer.

**DDAVP—Effectiveness Testing**

DDAVP was approved for use in the United States for individuals with mild hemophilia A and von Willebrand’s disease in 1983. DDAVP is a synthetic antidiuretic hormone that has been shown to release Factor VIII from endothelial tissue. If more Factor VIII is present in the bloodstream before an injury, the resulting hemorrhage will not cause as much damage as it might have. Unfortunately, the response to DDAVP is not universal. Personnel at hemophilia treatment centers test baseline levels of Factor VIII, administer DDAVP, and then reassess circulating levels of Factor VIII to determine the effectiveness of DDAVP. DDAVP treatment is considered effective if Factor VIII levels increase three- to five-fold. If this is the case, DDAVP can then be used to prevent and treat athletic injuries.

Two major factors that might influence the use of DDAVP are the expense of the product and convenience. One bottle of DDAVP nasal spray costs $149 and contains 25 doses. Each application would cost approximately $6, and the bottle would last about a month. The expense incurred will be quite high if the university’s or individual’s insurance policies do not cover prescription medication of this nature. Another influence with regard to the use of DDAVP is that plasma Factor VIII peaks approximately 1.5 hr after DDAVP use and generally keeps Factor VIII levels up for about 8 hr. The athlete would have to remember to administer the DDAVP before competition, which might be inconvenient. In addition, with repeated exposures administered every 12–24 hr, there is a diminished effect of the drug. This can be reversed by suspending treatment for 1–6 weeks, depending on the individual. A final influence on DDAVP use is the possible side effects. Facial flushing, transient headaches, nausea, nasal congestion, runny nose, and abdominal cramps are among the potential side effects, although they are rarely reported in clinical trials.

**Educating Sports-Medicine Staff**

**General Knowledge of Hemophilia and Injury Management**

Managing injuries in athletes with hemophilia is not common in competitive athletics. When a medical staff is accountable for the welfare of athletes with special needs, an individualized management plan is warranted. The athletic trainer must recognize the symptoms associated with deficiencies in coagulation and determine when to enact the emergency management plan, which includes the use of prophylactic medications and infusing Factor VIII. An important part of the injury-management process involves communication with the athlete. An athlete with hemophilia will be able to recognize when the inflammatory response to an acute trauma is associated with hemorrhaging beyond the normal response.

**Individual Management Plan**

A well-constructed emergency action plan should already account for potential medical emergencies including bleeding conditions. When a medical staff is aware of an athlete’s particular medical condition, it is the physician’s and athletic trainer’s responsibility to create a management plan that accounts for bleeding emergencies. The individual management plan should be a collaborative effort of the medical staff and the athlete, with consideration of specific management criteria documented and placed in the athlete’s medical file. It should include the responsibilities each participant will assume in the event of a bleeding emergency. It is important that athletes with hemophilia recognize that their role in the management plan is essential. The risks will essentially be elevated if an athlete with hemophilia does not recognize the potential threat associated with certain sport activities.

**Home-Contest Considerations**

During home contests, it is essential that the medical staff be prepared to manage the injuries of athletes with bleeding disorders. In addition to a well-established emergency action plan, DDAVP should be
made available if it has been shown to be effective in an athlete’s blood coagulation. If DDAVP is used as a prophylactic measure, the athlete must remember to initiate treatment before competition. In the event of a bleeding emergency, a unit of Factor VIII should be available on-site. In competitive events that sponsor an on-site team physician, the Factor VIII could be infused promptly, minimizing the severity of blood loss. Most often, these medical emergencies require physician care at an emergency room, and the Factor VIII and DDAVP should accompany the athlete in case the medical facility is not prepared for such an emergency and none is immediately available.

Away-Contest Considerations

When traveling to away contests, an athlete with hemophilia poses an added concern for the athletic trainer. The DDAVP and Factor VIII need to be transported in a special vessel. The DDAVP can be stored at 22 °C, controlled room temperature. This is convenient, but DDAVP lasts longer if refrigerated. If the athlete participates in an outdoor sport, temperatures might vary. This would require a storage vessel that can maintain room temperature. In addition to transporting DDAVP, the sports-medicine staff should also carry rFVIII to be used for treatment in the event of an injury. More critical during away contests is the need to obtain directions and contact numbers for the nearest hemophilia treatment center. Many emergency rooms are not equipped to handle hemophilia-related bleeding emergencies, and oftentimes transport to the closest hospital will in fact delay the management of injury.

Conclusion

There are many benefits of physical activity for the general population. Improved cardiorespiratory efficiency, control of body fat, improved strength and flexibility, and improved psychological and emotional well-being are among these. The same benefits from exercise, as well as some additional benefits, can be obtained by individuals with hemophilia. Muscle power and good range of motion can be maintained through exercise, reducing the number and severity of bleeding episodes caused by abnormal stresses. In addition, exercise has also been shown to increase the level of circulating FVIII. The benefits of exercise support the importance of having opportunities for individuals with hemophilia to participate in athletics. Athletic trainers and other sports-medicine professionals must recognize the special medical needs associated with caring for athletes who have hemophilia. In addition, they must establish an individual injury-management plan based on considerations relevant to the care of all athletes with hemophilia.

References


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