What is known about care of a newborn with hemophilia?

Decker, K , Nagel, K, Chan, AK.

Department of Pediatrics, McMaster University/ Hamilton-Niagara Regional Hemophilia Program/ McMaster Children's Hospital/ Hamilton Health Sciences/ Hamilton, Ontario, Canada

Introduction

•Hemophilia is a life long bleeding disorder that prevents blood from clotting properly.[1] Hemophilia A (factor VIII deficiency or classic hemophilia) and hemophilia B (factor IX deficiency or Christmas disease) are the most common types of hemophilia.[2]

• Hemophilia is primarily a hereditary blood disorder inherited as X-linked recessive traits which means hemophilia mostly affects males.[2]

• Carrier mothers can pass hemophilia to their sons on the X chromosome, an ultrasound can be performed to determine fetal gender. However, 30% of cases occur as a spontaneous mutation. In these cases, there is no family history of hemophilia.[3]

•In a newborn, this disorder presents a plethora of challenges, initially with diagnosis and management due to the unique features of this age group.[4]

•Management requires an inter-professional collaboration between three specific services. These services are obstetrics, hematology and neonatology.[4]

•Management is even more complicated where a family history is absent however when a hemophilia carrier is known, a diagnosis of hemophilia A can be confirmed at birth from a cord blood sample.[5]

•While major bleeding is relatively uncommon during this period, neonates are susceptible to intracranial hemorrhage related to birth trauma.[5]

• During the first few days of life the incidence is higher than any other stage of childhood.[5] In 1999, Buchanan et al proposed the use of prophylactic factor VIII during the first 24 hours following birth for the prevention of intracranial hemorrhage.[6]

• In the ensuing years this has been challenged because the incidence of inhibitor development maybe associated with early dosing of FVIII.

•Evidence based guidelines for care of a neonate with hemophilia have been published[7], but several studies have found that practice across institutions, are inconsistent even within institutions involving these practice guidelines.[4,5,8]

Methods

•This is a retrospective chart review and included all severe hemophilia patients born at McMaster Children's Hospital, Ontario, Canada for the last ten years.

•Data collected included: date of birth, birth weight, type and number of blood draws, invasive interventions (assisted birth, heel pokes, intravenous use, IM injections)and bleeding complications.

Results

•A total of nine charts were reviewed.

•All births occurred in the hospital.

• Birth weight ranged from 2687 to 4195 grams with a mean of 3437 grams.

•Type of blood draws included heel pokes, peripheral IV (PIV) and cord blood.

• Invasive interventions included heel pokes (n=28), PIV (n=2), and vitamin K injections (n=9).

•All babies were born by vaginal delivery.

•Two patients had an unknown family history of hemophilia.

•Two patients experienced bleeding complications requiring administration of factor VIII; one due to bleeding post heel poke, and the second for cephalohematoma and anemia. Both of these patients had a known family history of hemophilia. A third patient required admission to NICU to monitor hypothermia, hypoglycemia, and cephalohematoma, but did not receive FVIII.

•None of the infants had any complications from their vitamin K injection.

Conclusion

•We have adapted our practice to include placement of topical thrombin at the bedside for use if infants experience bleeding from heel pokes.

•A summit meeting with nursing leaders from hematology/hemophilia, neonatology and obstetrics to clearly define practice guidelines at McMaster Children's Hospital is needed.

References

World Federation of Hemophilia. (2006). *What is Hemophilia?* Retrieved October 25, 2010 from http://www.wfh.org/index.asp?lang=EN

Springhouse. (2009). *Evidence-Based Nursing Guide to Disease Management*. Philadelphia, PA: Lippincott Williams & Wilkins.

Price, V. E., Hawes, S.A. & Chan, A.K.C. (2007). A practical approach to hemophilia care in children. *Paediatrics & Child Health*, 12(5), 381-383.

Chalmers E.A. Haemophilia and the newborn. Blood Reviews. 18 (2) (pp 85-92), 2004.

Chalmers E.A., Williams M.D., Richards M., Brown S.A., Liesner R., Thomas A., Vidler V., Pasi K.J., Hill F.G.H. Management of neonates with inherited bleeding disorders – A survey of current UK practice [1]. Haemophilia. 11 (2) (pp 186-187), 2005.

Buchanan G.R. Factor concentrate prophylaxis for neonates with hemophilia. Journal of Pediatric Hematology/Oncology. 21 (4) (pp 254-256), 1999.

Chalmers E.A., Williams M., Brennand J., Liesner R., Collins P., Richards M. Guideline on the management of haemophilia in the fetus and neonate. British Journal of Haematology. 154 (2) (pp 208-215), 2011.

Chan A.K.C., Decker K., Warner M. Adherence with haemophilia treatments: A survey of haemophilia healthcare professionals in Canada. Haemophilia. 17 (5) (pp 816-817), 2011.

WFH2014 200-P-W

Inter-professional care

Kay Decker

Poster presented at: WFH 2014 World Congress Melbourne, Australia July 11-15

PosterSessionOnline.com