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ORIGINAL ARTICLE

Review of patients studied for coagulopathy in a haematology/oncology unit☆

I. Romero[a], N. Conde[a,*], D.G. Aldana[a], A. Ruano[b], A. Fernández-Teijeiro[a]

[a] *Unidad de Onco-Hematología Pediátrica, Hospital Universitario Virgen Macarena, Unidad de Gestión Clínica Intercentros de Oncología Pediátrica, Hospitales Universitarios Virgen Macarena y Virgen del Rocío, Sevilla, Spain*
[b] *Sección de Trombología, Hospital Universitario Virgen Macarena, Unidad de Gestión Clínica Intercentros de Hematología, Hospitales Universitarios Virgen Macarena y Virgen del Rocío, Sevilla, Spain*

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Paediatric bleeding;
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Abstract
Introduction: Symptoms/signs suggestive of coagulopathy is a frequent complaint in Paediatric Haematology units. Both the clinical and family history are essential for diagnosis.
Patients and methods: Retrospective and descriptive study of patients referred to a Paediatric Haematology unit of a tertiary hospital for possible coagulopathy during 2012.
Results: A total of 47 children were studied, of whom 61.7% had not previously presented bleeding. The most frequent reason for referral was a prolonged activated partial thromboplastin time without any haemorrhage (42.5%), of these, 25% were diagnosed with coagulopathy with a real risk of bleeding. Patients referred due to a prolonged activated partial thromboplastin time with bleeding more frequently (41.7%) present coagulopathy with a real risk of bleeding. Children with a family history of bleeding are diagnosed more frequently with coagulopathy with real risk of bleeding: 37.5% (family history) *vs.* 14.3% (no family history). The most frequent diagnoses were: healthy children (48.9%), von Willebrand type 1 disease (19.1%), factor XII deficiency (19.1%), factor XI deficiency (4.2%), prekalikrein/high molecular weight kininogen deficiency (2.1%), acquired deficiency of factor X (2.1%), and factor IX deficiency (2.1%).
Conclusions: A thorough personal and family bleeding history and physical examination are the first steps for a correct differential diagnosis. The reason for referral should be based more on clinical bleeding and not just on an abnormal coagulation time. The most frequent diagnoses were type 1 von Willebrand disease and factor XII deficiency.

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* Corresponding author.
E-mail address: nuriaconde@hotmail.com (N. Conde).

PALABRAS CLAVE
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Revisión de los pacientes estudiados por coagulopatía en una unidad de Oncohematología

Resumen
Introducción: Los síntomas/signos indicativos de una coagulopatía son un motivo de consulta frecuente en las unidades de Hematología Pediátrica. Tanto la clínica como los antecedentes familiares son fundamentales para el diagnóstico.
Pacientes y métodos: Estudio retrospectivo y descriptivo de los pacientes derivados a una consulta de Hematología Pediátrica de un hospital de tercer nivel por posible coagulopatía durante el año 2012.
Resultados: Se estudiaron 47 niños. El 61,7% no había presentado previamente sangrado. El motivo de derivación más frecuente fue un tiempo de tromboplastina parcial activada alargado sin hemorragia (42,5%); de estos, un 25% fue diagnosticado de una coagulopatía con riesgo real de sangrado. En los pacientes derivados por tiempo de tromboplastina parcial activada alargado con clínica hemorrágica se detecta una coagulopatía con riesgo real de sangrado con mayor frecuencia (41,7%). En los niños con antecedentes familiares de sangrado se diagnostica con más frecuencia una coagulopatía con riesgo real de sangrado: 37,5 vs. 14,3% (niños sin antecedentes familiares). Los diagnósticos han sido: sano (48,9%), enfermedad de von Willebrand tipo1 (19,1%), déficit de factor XII (19,1%), déficit de factor XI (4,2%), déficit de precalicreína/cininógeno de alto peso molecular (2,1%), déficit adquirido de factor X (2,1%) y déficit de factor IX (2,1%).
Conclusiones: Los antecedentes personales y familiares de sangrado orientan el diagnóstico de una coagulopatía. El motivo de derivación debería basarse en mayor medida en la clínica hemorrágica y no solo en un tiempo de laboratorio alterado. Los diagnósticos más frecuentes han sido enfermedad de von Willebrand tipo 1 y déficit de factor XII.

Introduction

Coagulopathy-indicative signs are a frequent reason for consultation in haematology. Its diagnosis can be difficult, due to specific bleeding age patterns: epistaxis (39%) and easy bruising (24%) are common without any haemorrhagic disorder. However, these can be the only clinical symptoms in children with coagulopathies who have not undergone any haemostatic challenge.[1]

In addition, differential bleeding/bruising diagnosis in children includes other entities, such as platelet alterations, vasculopathies and abuse.

Among hereditary coagulopathies, the most frequent is von Willebrand disease (vWD), which has a prevalence of approximately 1%, according to studies.[2,3] More recent studies, however, have reported a 0.1% prevalence of clinically symptomatic vWD in children.[4] This is followed by factor VIII (haemophilia A) and factor IX (haemophilia B) deficiency, with a predominance of 1:5000 males and 1:30,000 males, respectively.[5]

Some factor deficiencies do not result in bleeding, such as the deficiency of some contact factors: high molecular weight kininogen (HMWK), prekallikrein and factor XII. These factors are not involved in the propagation phase of the coagulation cascade and, therefore, their deficiency, in spite of prolonging the activated partial thromboplastin time (APTT) *in vitro*, is not associated with bleeding *in vivo*.[6]

In this study, we set out to determine which services refer the greatest number of patients for coagulation analysis and the reason for referral, to correlate laboratory findings with clinical symptoms of bleeding, and to determine the most common diagnoses made in our hospital. Greater insight into these patients will help improve diagnosis protocols.

Patients and methods

Retrospective descriptive analysis of children under study due to suspected coagulopathy at the paediatric oncohaematology unit of the Virgen Macarena University Hospital (UH) in Seville in 2012.

At this unit, in 2012, all paediatric solid/haematological tumours (except for brain tumours) and non-oncologic haematology disorders were studied and treated. Patients diagnosed with haemophilia A or B were referred to the Haemophilia Unit from the Rocio UH in Seville after diagnosis.

There were 20 new oncology patients in 2012, and 120 haematological patients.

Clinical treatment was given by paediatric haematologists and laboratory techniques by haematologists.

We have taken into consideration children with altered coagulation times, those with normal coagulation and bleeding times (after ruling out thrombocytopenia), and those studied due to having a family member with a hereditary