

## Congenital bleeding disorders: When to evoke in neonates?

**Sadok Hannachi\***, Hatem Ben Salem and Sonia Blibech

University of Tunis El Manar, Tunisia

**Introduction:** Congenital Bleeding Disorders (CBD) comprises a heterogeneous group of diseases that reflect abnormalities of blood vessels, coagulation proteins and platelets. Patients with rare CBD may have a broad spectrum of clinical symptoms, ranging from mucocutaneous bleeding to life-threatening hemorrhages, such as those occurring in the central nervous system.

**Methods:** A retrospective descriptive study of all neonatal cases of CBD registered in the Neonatal Intensive Care Unit of the Military Hospital of Tunis, through the 20 previous years.

**Results:** The first observation was about a full term male infant, with no significant familial history, who declared at 4 hours of life an overwhelming hemorrhage. Initial blood tests revealed biologic signs of disseminated intravascular coagulation. The assays of IX, VIII and Von Willebrand factors were normal. Brain MRI showed an intraventricular hemorrhage with quadriventricular hydrocephalus. Analysis of the plasma revealed a severe deficiency of ADAMTS13. The patient has since required periodic prophylactic Fresh Frozen Plasma (FFP) infusions. With a decline of seven years, he kept moderate encephalopathy. Genetic study confirmed the diagnosis of Upshaw-Schulman syndrome.

The second case was about a newborn with an enormous caput succedaneum. The blood count was consistent with severe anemia and coagulation studies revealed a prolonged Activated Partial Thromboplastin Time (APTT). Coagulation factors were measured, with a very low factor VIII activity (<1%) which was compatible with severe type-A hemophilia. Despite introducing the specific treatment, the infant died at the age of 18 months due to a severe hemorrhagic episode.

The third and fourth cases were about a brother and his sister who presented an umbilical hemorrhage that wasn't initially explored. Both of them have had seizures due to an intracranial hypertension and multiple episodes of bruising. The blood tests showed no anomalies aside from a low rate of XIII factor.

**Conclusion:** In our patients, the onset of CBD was originally declared with a severe neonatal hemorrhagic syndrome. Indeed, early diagnosis is mandatory to adequately monitor the disease, prevent organ damage and severe sequelae.

## Biography

Sadok Hannachi is affiliated to University of Tunis El Manar, Universitaire Farhat Hached, Tunis 1068, Tunisia

---

**Received:** March 17, 2023; **Accepted:** March 19, 2023; **Published:** July 17, 2023

---