

# Severe neonatal subgaleal hemorrhage as the first presentation of hemophilia A

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## SUMMARY

**Introduction** Subgaleal hemorrhage is a rare but potentially fatal birth trauma. It is caused by rupture of the emissary veins (connections between the dural sinuses and scalp veins), followed by the accumulation of blood between the epicranial aponeurosis and the periosteum. Usually, it is associated with instrumental delivery (vacuum extraction, forceps delivery), but it may also occur spontaneously, suggesting the possibility of congenital bleeding disorder.

**Case Outline** A full term male neonate was born at 40 weeks gestation by spontaneous vaginal delivery, with birth weight of 3,700 g. The Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. At the age of 23 hours, the baby became pale and lethargic. Large fluctuant swelling on his head was noted. He developed severe anemia and hypovolemia as a result of massive subgaleal hemorrhage. After successful treatment, the baby fully recovered. Follow-up and further evaluation revealed hemophilia A as a result of a *de novo* mutation.

**Conclusion** This case illustrates that subgaleal hemorrhage may be the first presentation of hemophilia A. Infants without obvious risk factors for developing subgaleal hemorrhage should be evaluated for congenital bleeding disorder. Successful outcome in affected infants requires early diagnosis, careful monitoring and prompt treatment.

**Keywords:** subgaleal hemorrhage; neonate; hemophilia A

## INTRODUCTION

Subgaleal hemorrhage (SGH) is an uncommon, potentially lethal, but often underdiagnosed condition in newborns. The prevalence of subgaleal hemorrhages is 0.04% to 0.15% of all deliveries, with mortality range from 12% to 25% [1, 2]. It is caused by the rupture of the emissary veins (connections between the dural sinuses and scalp veins), followed by separation of the epicranial aponeurosis from the underlying periosteum. This can create a compartment in which approximately 250 ml of blood could be accumulated [1]. SGH usually occurs as a complication of instrumental delivery (vacuum extraction or forceps). This can lead to the loss of up to 50–75% of blood volume, resulting in shock, anemia, consumptive coagulopathy, hyperbilirubinemia, encephalopathy, and possibly death.

## CASE REPORT

We report a case of a male neonate with massive subgaleal hemorrhage. This full term infant was born to non-consanguineous and healthy parents. The pregnancy had been uneventful. Delivery was spontaneous, vaginal, and without complications, at 40 weeks of gestation. Birth weight was 3,700 g (50 perc), length 52 cm (50 perc), and head circumference 34 cm (25 perc). The Apgar scores were 9 and 10 at one and five minutes, respectively.

The baby received a prophylactic intramuscular dose of vitamin K. Postpartum course was uneventful until the 23rd hour of life when the baby suddenly became pale and lethargic with diffuse swelling of the scalp. The working diagnosis of subgaleal hemorrhage was made, parenteral antibiotics and hydration were started, and the baby was transferred to a tertiary center neonatal intensive care unit (NICU) for further management.

Physical examination on admission showed a severely ill, pale baby with weak painful cry and decreased muscle tone. Vital signs were as follows: heart rate 180 bpm, respiratory rate 64 bpm, blood pressure 59/37 mmHg, oxygen saturation 97% (room air), capillary refill time 5 s. There was a fluctuant edema of the scalp, eyelids, and forehead, with downward displacement of the earlobes. Head circumference was 38.5 cm (Figure 1).

Laboratory investigation revealed an uncompensated metabolic acidosis (pH 6.99, pCO<sub>2</sub> 6.74 kPa, pO<sub>2</sub> 5.13 kPa, St. HCO<sub>3</sub> 8 mmol/l, BE -22.5 mmol/l, severe anemia (Hgb 30 g/l, hematocrit 10%, RBC 0.94 T/l), thrombocytopenia (110 G/l), and pathological coagulation parameters (PT 26 s, INR 2.38, APTT 41.3 s, fibrinogen 1.9 g/l). The baby was immediately endotracheally intubated and mechanical ventilation was started. Hypovolemia was initially treated with boluses of normal saline. Over the first 12 hours the baby received repeated transfusions of packed red cells (a total of 30 ml/kg) and fresh frozen plasma (15 ml/kg). The

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**Figure 1.** Head swelling with marked periorbital and periauricular edema in a newborn with subgaleal hemorrhage (parental consent was obtained for the publication of this photograph)



**Figure 2.** A head CT reveals cerebral edema and massive subgaleal hemorrhage

administration of blood products led to significant improvement (Hgb increased to 102 g/l) with no signs of continued bleeding. Early after admission the baby had tonic seizures treated with phenobarbitone. After stabilization, further investigations were performed. Cranial ultrasound showed edema of subcutaneous head tissue with diffuse hematoma (largest diameter of 13 mm) and no evidence of intracranial bleeding. Head CT confirmed severe subgaleal hemorrhage (largest diameter of 17 mm), minimal frontal epidural hematoma associated with brain edema, without signs of intraventricular or parenchymal bleeding (Figure 2).

A diagnosis of hemorrhagic shock secondary to subgaleal hemorrhage was made. A neurosurgical consultation was obtained, and conservative treatment was recommended. Pathological coagulation profile on admission was initially assigned to a consumptive coagulopathy. By the age of three days the baby was extubated, without any signs of neurological impairment. However, repeated coagulation profiles revealed persistently prolonged values of activated partial thromboplastin time (aPTT) (R 1.947 ... 2.753 ... 2.360 ... 2.510). Further hemostasis analyses were

performed, showing decreased levels of factor VIII (FVIII 7%) and the working diagnosis of hemophilia A was made. The infant was discharged home at the age of three weeks postnatally. Follow-up visits with pediatric hematologist were scheduled and further evaluation led to definitive diagnosis of hemophilia A at the age of six months, as a result of a *de novo* mutation.

## DISCUSSION

SGH occurs more commonly in vacuum- and forceps-assisted deliveries, although there are reports of SGH in spontaneous vaginal deliveries associated with macrosomia, fetal distress, prolonged second stage of labor, prematurity, primigravidity, and precipitous delivery [3]. The incidence of SGH drops markedly to 4/10,000 in spontaneous vaginal deliveries and is rarely reported in non-traumatic cesarean sections [1].

The coagulation system of the newborn is immature, with lower levels of most of the coagulation factors. However, the factor VIII level is normal in newborns. Hemophilia A and B, both inherited as X-linked recessive disorders, are the most common inherited bleeding disorders to present in the newborn period. At least one third of all hemophilia cases occur without a positive family history. Intra- and extracranial hemorrhage are major causes of morbidity and mortality in newborns with hemophilia [4, 5]. The overall incidence of ICH in individuals with hemophilia regardless of age is 2.2–7.5% and approximately 50% of cases occur in the newborn period. The cumulative incidence of cranial hemorrhage (intra- and extracranial) in a retrospective study was 3.8% [6]. However, the true incidence of SGH in newborns with hemophilia remains unknown, mostly because of rare reporting of cases and the fact that SGH and cephalhematoma cases are usually combined into a single category of extracranial hemorrhage [1].

Our patient was born at term, from uneventful pregnancy with unremarkable family history and without apparent risk factors for SGH. From the moment the head swelling was noted, his condition worsened rapidly, with signs of severe anemia and hypovolemic shock leading to the admission in the tertiary NICU. Aggressive volume resuscitation and packed red cell transfusions were the mainstay of therapy leading to a full recovery. The SGH diagnosis was clinical with latter confirmation by head CT scan. The pathological coagulation profile was initially attributed to consumptive coagulopathy. However, after clinical improvement, persistently prolonged aPTT suggested the possibility of congenital bleeding disorder. Decreased level of factor VIII was revealed (FVIII 7%), and the diagnosis of mild form of hemophilia A due to a *de novo* mutation was made subsequently.

Our case illustrates that despite a non-traumatic delivery, SGH should also be considered as a differential diagnosis in infants with anemia, severe acidosis, and shock. Also, if SGH occurs in newborns without usual perinatal risk factors, firstly vacuum and forceps assisted deliveries, an evaluation for bleeding disorders should be performed.

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## Масивно субгалеално крварење као прва презентација хемофилије А код новорођенчета

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### КРАТАК САДРЖАЈ

**Увод** Субгалеално крварење је ретка, потенцијално животну угрожавајућа порођајна повреда. Узроковано је руптуром емисарних вена које повезују синусе тврде моздане овојнице и вене поглавине са последичним накупљањем крви између епикранијалне апонеурозе и покоснице. Најчешће настаје као последица инструменталног порођаја (вакуум екстракција, порођај форцепсом), али може настати и спонтано, када може указати на урођени поремећај хемостазе.

**Приказ болесника** Мушко рочно новорођенче гестацијске старости 40 недеља рођено спонтаним вагиналним порођајем, порођајне телесне масе 3700 g, Апгар скор 9/10. У 23. сату живота новорођенче постаје бледо и летаргично. Регистрована је појава флукутирајућег едема поглавине.

Новорођенче развија знаке тешке анемије и хиповолемије као последице масивног субгалеалног крварења. Након примењене терапије долази до потпуног опоравка. Спроведеном евалуацијом и праћењем утврђено је да новорођенче болује од хемофилије А, која је резултат мутације *de novo*. **Закључак** Приказ случаја указује да субгалеално крварење може бити прва презентација хемофилије А. Новорођенчад са субгалеалним крварењем без јасних фактора ризика треба испитати у правцу постојања наследног поремећаја хемостазе. Погодан исход зависи од раног постављања дијагнозе, интензивног мониторинга и енергичног лечења пацијента.

**Кључне речи:** субгалеално крварење; новорођенче; хемофилија А

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