

# Congenital pemphigus syphiliticus: a characteristic feature of a forgotten disease

Sakviseth Bin 

Neonatal Intensive Care Unit,  
Calmette Hospital, Phnom Penh,  
Cambodia

**Correspondence to**  
Dr Sakviseth Bin;  
sakviseth\_bin@yahoo.com

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## DESCRIPTION

The preterm male infant was delivered at 30 3/7 weeks of gestational age by an emergency C-section due to placenta previa, with haemorrhage and fetal distress. The mother, 21 years old, gravida 2 parity 1, had no underlying diseases. The antenatal care was done regularly at a provincial health centre; syphilis and HIV were negative at the first prenatal visit in the first trimester. She was transferred to our tertiary centre for vaginal bleeding.

The birth weight was 1150 g (20th percentile), and the Apgar scores were 3/4/5 at 1, 5 and 10 min, respectively. Pressure positive ventilation was given, and he was intubated at 3 min of life and transferred to the neonatal intensive care unit. Umbilical cord venous pH was 7 with lactate of 10 mmol/L. Per unit protocol, septic screen was done, and empiric antibiotics were prescribed. Gastric aspirate and haemoculture (peripheral and from central line) were later shown to be negative. Initial blood tests showed a white blood cell count of  $28.3 \times 10^9/L$  with immature/total neutrophil ratio of 0.05, severe anaemia (haemoglobin 105 g/L), platelets of  $190 \times 10^9/L$  and procalcitonin of 0.92 ng/mL.

Due to the presence of hepatomegaly (3 cm below the costal margin) and pemphigus on both feet ([figure 1](#)), syphilis tests, including Rapid Plasma Reagin (RPR) and Treponema Pallidum Hemagglutination (TPHA), were requested:

- ▶ The mother's titre : RPR 1:4, TPHA 1:640
- ▶ The infant's titre: RPR 1:4, TPHA 1:640

The diagnosis of maternal and congenital syphilis was made, and the antibiotics were changed to benzylpenicillin at 3 hours of life. This case report illustrates a severe case of perinatal asphyxia, masking a congenital syphilis.



**Figure 1** Pemphigus syphiliticus.

## Learning points

- ▶ Congenital syphilis is severe but preventable with proper maternal screenings, leading to an early, timely and adequate treatment of maternal syphilis.
- ▶ Pemphigus syphiliticus is rare but pathognomonic, warranting a thorough physical examination of the newborn.
- ▶ The diagnosis tools and treatments are easily accessible and inexpensive in our economical settings.

Regretfully, the neonate died at 32 hours of life. He was on conventional ventilation for 12 hours (Synchronized Intermittent Mandatory Ventilation (SIMV)) and on High-Frequency Oscillation (HFO) for 20 hours. He got two transfusions of red blood cells.

Mother-to-child transmission of *Treponema pallidum* can occur during any stage of pregnancy. Fetal infection from untreated or inadequately treated maternal syphilis can lead to serious adverse birth outcomes, including fetal loss, stillbirth, early infant death, symptomatic infected newborns, premature delivery and low birth weight.<sup>1</sup>

Clinical features of early congenital syphilis are variable, and the neonates are mostly asymptomatic at birth. The most common manifestations are hepatomegaly, jaundice and skin rash; other symptoms include rhinitis, lymphadenopathies, central nervous invasion and bone involvement.<sup>2</sup> The most prominent cutaneous lesions are copper-coloured, maculopapular rash, mostly on the palms and soles. Pemphigus syphiliticus is pathognomonic but rare. It is characterised by fluid-filled bullae, which appear mostly at the extremities and tend to rapidly peel and form crust.<sup>3</sup>

The diagnosis of congenital syphilis depends on multiple factors such as maternal serology, adequacy of her treatment, physical examination of the newborn and paired serology using non-treponemal tests such as Rapid Plasma Reagin and Venereal Disease Research Laboratory. Penicillin G remains the most effective spirocheticidal drug of choice.<sup>4</sup>

Congenital syphilis can be prevented by early diagnosis of maternal syphilis through repeated screenings and on-time and adequate treatment. It was also shown that even incomplete maternal



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penicillin therapy could reduce the risks of severe neonatal complications.<sup>5</sup>

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#### ORCID iD

Sakviseth Bin <http://orcid.org/0000-0003-2513-8690>

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