

# Congenital varicella syndrome in a monozygotic diamniotic twin pregnancy

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Congenital varicella syndrome encompasses a broad spectrum of malformations present in children of mothers who developed chickenpox during the first 20 weeks of gestation. We report a case of a monozygotic diamniotic twin pregnancy, with maternal exposure to chickenpox during the thirteenth week of gestation, which produced one symptomatic and one healthy child.

**Key words:** Chickenpox, congenital varicella syndrome, diamniotic, herpes zoster virus, monozygotic

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## INTRODUCTION

Congenital varicella syndrome (CVS) is an uncommon complication of maternal varicella infection occurring in about 2% of the newborns of women infected with varicella between the seventh and the twenty-eighth weeks of pregnancy. Primary varicella infection during pregnancy can rarely result in intrauterine infection of the fetus causing congenital varicella syndrome.<sup>[1]</sup>

The typical clinical manifestations of congenital varicella syndrome include skin lesions in dermatomal distribution (73%), neurological defects (62%), ocular abnormalities (52%), limb hypoplasia (46%), and low birth weight (23%) among others minor defects.<sup>[2]</sup> We report the clinical features of the first case of congenital varicella in a monozygotic diamniotic twin pregnancy.

Case report in the University Hospital of Valle, Cali, Colombia. 2008.

A 24-year-old woman was gravida 1, para 0, with an early uneventful twin monozygotic diamniotic pregnancy and a tenth week ultrasound normally reported. In the thirteenth week of gestation the patient presented with fever for one week and pruritic vesicular eruptions on the skin, consistent with the classic presentation of chickenpox. The patient had no history of chickenpox in childhood and had not received immunization against it. No antiviral treatment was given. At 24 weeks of pregnancy the

patient underwent a new ultrasound in which one of the fetuses presented with decreased head circumference and facial abnormalities.

A Cesarean section was performed at 36 weeks of pregnancy. The first newborn was a 2,588 g, 45 cm long female, with Apgar scores of 2 and 0, at one and five minutes, respectively. It presented CVS stigmata, with a right hemicranial sloughing scalp, homolateral microphthalmia, microtia and hemifacial microsomia, and commissural labial deviation (Figures 1 and 2). The second newborn was a 2,750 g, 49 cm long female, with Apgar scores of 9 and 10, at one and five minutes, respectively, with no physical abnormalities.

At birth, the mother's and newborn's IgG varicella antibodies (by Enzyme-linked immunosorbent assay (ELISA)) were present at a titer of 13.3 U/mL and 9 U/mL, respectively. Both had negative specific IgM antibodies. There were no detectable antibody titers in the healthy newborn.

## DISCUSSION

There is a very high prevalence for herpes zoster virus (HZV) in the pediatric population; about 85% of people younger than 15 years of age have been exposed to the virus, and 90% of the child-bearing age women will be immune to the disease. Complications of varicella are more frequent and severe in adults than in children, and this is especially true during pregnancy, wherein, both the mother and the fetus are affected.<sup>[3]</sup> When primary

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**Figure 1:** Female neonate with hemangiomatous skin lesions compromising the right scalp and forehead, hemifacial microsomia, homolateral microphthalmia, microtia, and commissural labial deviation

infection with HZV occurs during gestation there is a chance of vertical transmission to the fetus at the point when the virus has colonized the mucous membranes.<sup>[4]</sup> There is a 25% risk of infection, and from these, 12% can result in CVS.<sup>[5]</sup>

The congenital varicella syndrome was first described by La Foret and Lynch, in 1947, and since then, approximately 130 cases have been reported. The estimated prevalence of CVS is one to five in 10,000 pregnancies, where primary infection in the first 20 weeks has been documented.<sup>[6]</sup> It has a 30% mortality and miscarriage rate of 3-8%.<sup>[7]</sup> The mechanism of CVS is thought to be a consequence of the reactivation of the HZV in the uterus, akin to the mechanism of HZ development, rather than being due to the primary infection.<sup>[8]</sup>

According to literature, the most common clinical features of CVS are: Skin lesions that resemble scars, burns or present with hemangiomatous characteristics with a dermatomal distribution (72%); neurological defects (62%) such as cortical atrophy, microcephaly, seizures and mental retardation; ocular alterations (52%) like chorioretinitis, cataract, Horner syndrome, and nystagmus; limb hypoplasia (46%); and low birth weight (23%), among other less common features.<sup>[9]</sup>

The patient in this report presented with hemangiomatous skin lesions compromising the right scalp and forehead, microphthalmia, microtia, and a right labial commissure deviation. Serological tests, clinical features, and the clear antecedent of varicella in the thirteenth week of pregnancy gave us reasons to establish a clinical and serological diagnosis. On the other hand, the lack of clinical manifestations in the other twin was remarkable, which suggested two possibilities: There was no vertical



**Figure 2:** Overview of the patient. Note that the commitment is confined exclusively to the face and skull

transmission of the infection or there was discordance in the response to exposition to the same agent. It was worth noting that the antibody titers in the fetus did not relate to the development of the syndrome, and that according to literature, there was no visible infection in 39% of the cases.<sup>[6]</sup>

In conclusion, the congenital varicella syndrome is an uncommon disease that has severe repercussions for both the mother and the fetus, which could be prevented by covering all the alternatives to prevent the infection, such as, specific and non-specific treatment and passive and active immunization, to decrease the risk of infection.<sup>[10]</sup> Thus, it must encompass a clinical assessment of the preconceptional and prenatal history in seronegative women, and exposure prevention of child-bearing age women without a history of HZV infection. In addition, further studies should be done on congenital varicella to understand the discordance among twins.

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