CASE STUDY

Hearing screening in a newborn with hyperbilirubinemia

Patient's details

Male infant born at 38 weeks of gestation from the third pregnancy of a 29-year-old mother, blood group O Negative, born by cesarean section, weighing 3,000 grams, length 48 cm, Apgar score 8/9 and born without complications. The infant was admitted to the hospital in his community at 36 hours of life with jaundice, with laboratory results indicating levels of indirect bilirubin (IB) at 38.8 mg/dL, direct bilirubin (DB) at 0.6 mg/dL and total bilirubin at 39.4 mg/dL, reticulocytes (Ret) at 25.6%, Hemoglobin (Hb) at 13.3mg/dL, hematocrit (Htc) at 38.4%, platelets (Plt) at 172,000 and blood type O positive. He was sent to the referral hospital to continue with clinical approach and treatment.

Initial evaluation and clinical diagnosis

Upon admission, he was identified as a newborn with jaundice, normal mental status and normal muscle tone. Initial laboratory results found increased levels of total bilirubin at 40.3 mg/dL, IB at 39.4 mg/dL, DB at 0.83 mg/dL, positive direct Coombs test and with a bilirubin/albumin ratio of 11.5 mg/g. At 40 hours of life, a plasma exchange was performed, which reduced bilirubin levels: TB at 21.7 mg/dL, DB at 0.43 mg/dL, IB at 21.3mg/dL. At 16 hours after plasma exchange, again an increase in bilirubin level was detected, with IB at 22.6mg/dL and Ret at 11.9%. The patient's mental status changed from alert to stupor with hypotonia and intensive phototherapy and intravenous immunoglobulin treatment was initiated.

Outcome

Clinical improvement was slow; after 7 days, the patient was found to be alert with weak suck action, reduced general movements and no change in muscle tone. Magnetic Resonance Imaging (MRI) of the brain was requested and hyperintensity was observed bilaterally in the globus pallidus in the T1-weighted sequence (Figure 1). A hearing screening evaluation was carried out using an otoacoustic emissions (OAE) test (Utilizing Echoscreen III – Natus Medical Incorporated), the results of which were PASS (Figure 2) for both ears (indicating integrity of the cochlear functioning of the ear). At the same time, an automated auditory brainstem response (aABR) test was carried out, the result of which was REFER bilaterally at 45 dB (Figure 3). Neurosensory hearing loss was suspected. To confirm the diagnosis, brainstem auditory evoked potentials (BEAPs) were requested, finding left ear with a threshold of 97dB and that, in the right ear, no response was evoked at the same auditory threshold (Figure 4). The audiological diagnosis was auditory neuropathy with deep bilateral neurosensorial hearing loss secondary to bilirubin toxicity. At 14 days of life, a new MRI was performed showing hyperintensity in the globus pallidus in the T2-weighted sequence (Figure 5 – see next page). These findings are typical of kernicterus: The neurological diagnosis at 10 months of life is dyskinetic motor disorder.

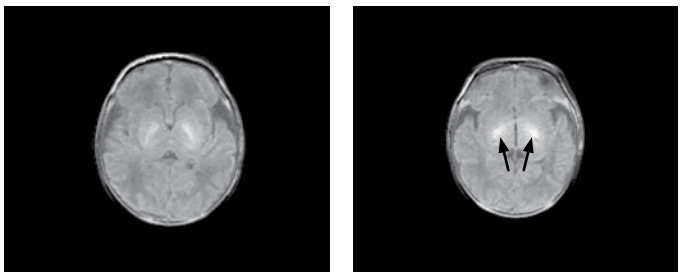

Figure 1: MRI T1-weighted sequence carried out at 7 days of life, presenting with bilateral hyperintensity in the globus pallidus (arrows), suggestive of bilirubin deposits.

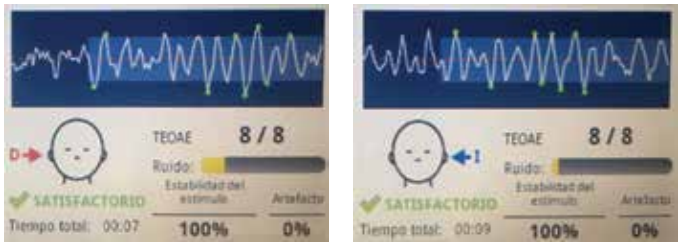

Figure 2: Otoacoustic emissions test with PASS results, proving the integrity of cochlear function in this patient.

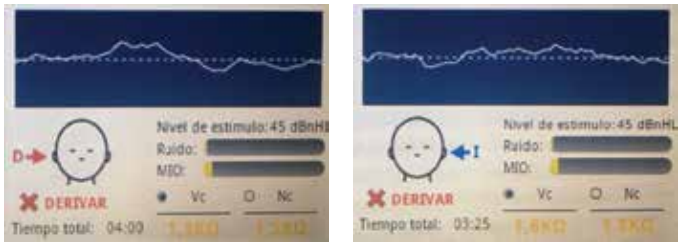

Figure 3: Automated auditory brainstem response with a REFER result (NOT SATISFACTORY) bilaterally with an auditory threshold of 45 dB.

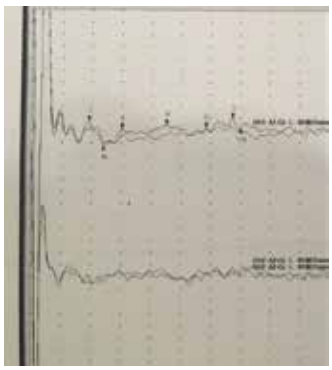

Figure 4: Brainstem auditory evoked potentials with an auditory threshold of 97 dB in the left ear, showing auditory response with waves I to VI. Absence of auditory response in the right ear.

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Discussion

Hyperbilirubinemia may affect 60 to 80% of newborns. Elevated levels of total serum bilirubin are generally mild, transient and in most cases inconsequential for children. However, kernicterus continues to be a significant issue worldwide. In developed countries, the prevalence of bilirubin-induced encephalopathy is 0.4 to 2.7 per 100,000 live births. In Latin America, 11.8 cases per 100,000 live births have been reported. Bilirubin-induced neurological dysfunction (BIND) is a clinical spectrum of neurological damage due to acute or sustained exposure of the central nervous system to bilirubin. Evidence has demonstrated the connection between hyperbilirubinemia and auditory system damage; these auditory effects can vary from subtle abnormalities in auditory processing and speech development to complete deafness. Hearing impairment is frequent in premature infants, while motor disorders are more common in full term infants, although this it is not a hard rule. All newborn infants with a history of hyperbilirubinemia should undergo complete auditory clinical evaluation.

Auditory Screening and further diagnostics tests is an important aspect of BIND-related hearing damage, with ABR being the preferred test for diagnosis. The auditory waves that are screened with this test are I (auditory nerve), III (cochlear nucleus) and V (lateral lemniscus). Auditory neuropathy spectrum disorder (ANSD) is characterized by abnormal auditory nerve function (abnormal or absent ABR waveforms) and presence of good cochlear function with normal otoacoustic emissions. There is growing evidence that patients with high levels of unconjugated (free) bilirubin experience worse neurodevelopmental outcomes and a higher incidence of hearing loss.

Up to 35% of babies with bilirubin levels greater than 20 mg/dL are subjected to transient or permanent ABR anomalies. However, a small proportion of newborns presenting with abnormal ABR findings may have serum bilirubin levels between 11 and 25 mg/dL.

In patients that required plasma exchange for clinical treatment, approximately 40% presented with abnormal findings; false positive rates were reported at less than 2%.

The auditory clinical approach for patients with a history of preterm birth and hyperbilirubinemia (the latter regardless of gestational age) should include OAE and ABR tests, which enable us to more comprehensively evaluate the auditory pathway to be able to establish a diagnosis and therapeutic plan.

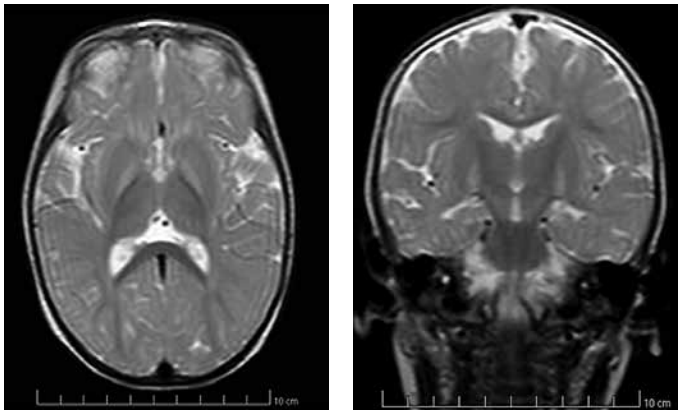

Figure 5: MRI T2-weighted sequence performed at 14 days of life with bilateral hyperintensity in the globus pallidus (arrows), confirming the diagnosis of chronic bilirubin-induced encephalopathy (kernicterus).

Bibliography

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