Tanınız Nedir?

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Our patient was a female neonate. She was delivered by C/S at 38 weeks of gestation to a 37 year old woman. The mother was followed irregularly during her pregnancy. The pregnancy was complicated by polyhydramnios determined in the last month. There was no history of parental consanguinity, infectious disease, hypertension, gestational diabetes, drug ingestion or X-ray exposure during the pregnancy. Her birth weight was 2250 gr (below 10 percentile according to New Ballard score). She was diagnosed **intrauterine growth retardation (IUGR)** and small for gestational age. Her length was 44 cms and head circumference was 33 cms. There was a history of feeding difficulty following the birth. She developed cyanosis after breast-feeding on day 3 and transferred to our hospital for further evaluation. Her physical examination revealed **peroral cyanosis, tachypnea, intercostal retractions**, **abnormal facial features** with **micrognathia, simple-low set ears, triangular facies and mouth, high-arched palate**. There was a 3/6º systolic murmur along the left sternal border. As the patient had neonatal pneumonia, she was applied mechanical ventilation plus antibiotic treatment.

Owing to patient's dysmorphis facies, we simply thought there might be another congenital anomalies. We tried to pass a firm catheter thorough each nostril into the nasopharynx to exclude choanal atresia, but we couldn't achieve. Rhinoscopy revealed bilateral **choanal atresia** and endoscopic dilatation was performed. **Tetralogy of Fallot (TOF)** was diagnosed by echocardiography.

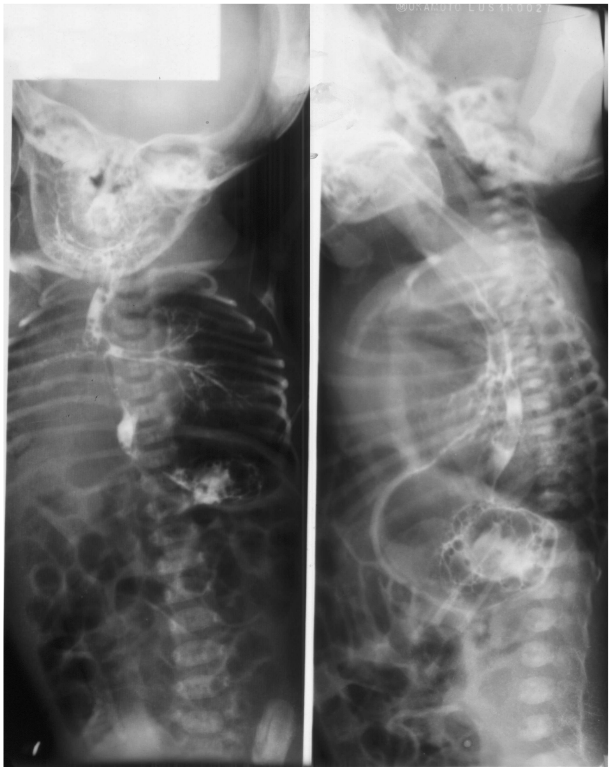

Figure 1. The barium swallow of the patient.

After extubation, we tried to feed the baby but respiratory distress was developed following

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the feeding. We performed barium swallow to exclude a tracheoesophageal fistula. The right bronchus, esophageus and stomach were filled with barium secondary to aspiration of barium (Figure 1). We suspected **velopharyngeal discoordination (probably dysfunction of 9/10th cranial nerves)**.

The patient's complete blood count, urine analysis and biochemichal studies were within the normal limits. Her cranial and abdominal ultrasonography, whole body skeletal graphics did not reveal any abnormality. Chromosomal analysis of the patient was normal female caryotype: 46;XX. The eye and fundus examinations of our patient were also found to be normal. We could not success extubating the patient because of recurrent aspiration pneumonia. As the patient developed neonatal cholestasis, we could not achieve feeding the baby with parenteral nutrition, we also could not achieve enteral nutrition secondary to velopharyngeal discordans. She developed **postnatal growth retardation,** and weighed 1650 gr postnatal 49th day. She died secondary to a severe aspiration pneumonia on day 50.

What is your diagnosis of our patient?