

Pregnancy And Delivery With Noonan's Syndrome: A Case Report

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ABSTRACT

A 31 year old woman (known case of Noonan's syndrome with cardiac disease) presented with a singleton pregnancy at 26 weeks. She was delivered uneventfully.

Keywords: Noonan's syndrome, atrial septal defect, oligohydramnios.

This 31 year old primigravid mother presented to us for her first antenatal checkup in the outpatient department, carrying a singleton gestation of 26 weeks. She was unmarried, and a known case of Noonan's syndrome, having undergone intracardiac surgery for atrial septal defect (ASD) with pulmonary stenosis (PS) earlier, but still having a residual ostium secundum left to right ASD of 8mm. She had mild tricuspid regurgitation and right bundle branch block, with an ejection fraction of 63%. She was on levothyroxine (50mcg) replacement therapy for gestational hypothyroidism. Ultrasound revealed a singleton foetus in cephalic presentation, with oligohydramnios (amniotic fluid index = 4), normal foetoplacental Doppler patterns, and no foetal anomaly on earlier scans. Foetal echocardiography was normal.

At 32 weeks, she presented in the emergency with gradual onset dyspnoea. She was admitted in the critical care unit (CCU), and received supportive management in the form of appropriate antibiotics, nebulisation, oxygen support and close monitoring, including paediatric cardiology input and foetal

surveillance via ultrasound and cardiotocography. The diagnosis was a mild lower respiratory tract infection, and she was discharged after 1 week in a stable condition.

At 35 weeks 4 days of gestation, she was admitted under us for safe confinement. All necessary investigations were done, and a detailed pre-anaesthetic evaluation and cardiological review were undertaken. Ultrasound showed a single live foetus at 33 weeks 3 days; cephalic, average liquor, upper anterior placenta, 2 loops of nuchal cord and normal Doppler studies. The modality of delivery was decided as elective Caesarean section, in view of foetal growth restriction with nuchal cord, and maternal cardiac condition. Antenatal steroids (betamethasone in standard 12mg doses) were administered 48 hours before surgery. Anaesthesia was provided as a single shot spinal in the L3-L4 interspace, using 2.2ml of 0.5% bupivacaine.

A Pfannenstiel incision and utilization of the Joel Cohen method during Caesarean were the cornerstones of our surgical technique. Low transverse incision was made over the uterus. She delivered a healthy baby girl of weight 1862g. Uterine closure was done in 2 layers,

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and the abdomen closed after securing haemostasis and matching counts, with subcuticular suturing to skin.

Post-operatively, she was shifted to the CCU for close monitoring. Post-surgical recovery was uneventful, and she was shifted to the ward after 24 hours of monitoring. Ultimately, she was discharged after 72 hours of surgery in a stable condition. The remainder of her post-partum period was uneventful.

Ethics Approval: As this was purely a case report without any intervention or analysis whatsoever, there was no research protocol involved. Consequently, Ethics Committee approval was not required.

Informed Consent: Written informed consent was obtained from the subject of this case report before the study began.

Author Contributions: Data acquisition, drafting and reviewing the work critically, final approval and accountability for all aspects of the work were shared equally between both authors.

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