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Case report

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## **58 days old infant TAR SYNDROME with COVID-19 infection**

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### **Abstract**

Thrombocytopenia absent radius is a rare congenital disorder which can be the most severe in the neonatal period. (1)

This disease is associated with absence (aplasia) of long thin bones of forearm; Thrombocytopenia and short stature. This disease might also be accompanied with structural alterations in Cardiovascular or Renal system. The patients also exhibits cow's milk intolerance.

Tar syndrome is an autosomal recessive pattern caused by a genetic disorder primarily due to the deletion of RBM8A gene. (2) The prevalence of this disease is between 0.5 -1:100000 and 1:240000 births affecting both the sexes equally.(3) Thrombocytopenia usually develops in the first week of life to months and usually decreases with age.

Tar syndrome patient coming for a procedure becomes an anaesthetic nightmare and in an infant subsequently diagnosed with Covid 19 is going to be a challenge for the anaesthesiologists. We report the case of a 58 days old infant diagnosed with Tar syndrome and with Covid 19 presented for central line insertion.

**Keywords:** Cardiovascular ;aplasia; anaesthetic

## **Introduction**

Thrombocytopenia absent radius syndrome is an extremely rare disorder and very less cases has been reported in India which has a multi organ association. Thrombocytopenia affects most number of patients in the initial months<sup>(3)</sup> due to the impaired bone marrow production of platelets.<sup>(4)</sup>The risk of bleeding might not correlate to the platelets counts. An upper and lower extremities anomaly along with cardiac anomalies such as Tetralogy of fallot, ASD and VSD is also another common manifestation in this subset of patients. The management of this scenario in a Congenital disorder affected with covid 19 has been a major hindrance. Due to bleeding diathesis, high risk of bleeding is expected and Covid 19 pandemic posed an additional unprecedented challenge to us with more precautions to be dealt with.

Covid 19 is a dysregulated inflammatory response <sup>(7)</sup> and administering general anaesthesia to infants can be a challenge as the anaesthetic drugs further suppresses immunity posing another great dilemma. To the best of our knowledge this is the first reported case of an infant diagnosed with TAR syndrome also affected with Covid 19 and has to undergo a procedure accompanied with general anaesthesia.

## **Case report**

58 days old infant, born in non-consanguineous marriage presented with the complaints of rectal bleeding with a haemoglobin drop from 9 gram/dl to 5 gram/dl and platelets decreasing from 35000 to 20000. The infant had undergone previous cbc test done outside which showed a 9 gram hemoglobin and in our Initial blood test results in our hospital revealed blood count:

## **Investigations**

Hemoglobin of 5gram/dl; Total leukocytes count (TLC) of 12500/uL, with Differential count revealing 49% Neutrophils, 39% Lmphocytes, Eosinophil and Monocytes 1% each, Platelet count, 20,000/uL; Coagulation profile – Prothrombin time, 14 sec (control 13 sec); partial thromboplastin time, 30 sec (control 30).Liver function test had the following results: Total serum bilirubin 1.2 mg/dL; Indirect serum bilirubin 0.8 mg/dL (direct), 0.4 mg/dL; and (SGPT) 28 U/L and (SGOT) 64U/L.

## **Treatment**

On the basis of clinical presentation, physical findings, isolated thrombocytopenia, the diagnosis of TAR syndrome was made and further evaluation it was also found that the child was also Covid rapid antigen positive with a small muscular ventricular septal defect. On admission it was found the airway & breathing was adequate and attaining room air saturation of 100%. To administer blood and blood products it was much required to access a peripheral line, which proved a challenge due to the Tar syndrome and deformities in upper limbs.The child was kept nil per oral and shifted to OR. Limited personals were included for the administration of anaesthesia care and specialised Personal Protection

equipment's was utilised. Basic monitors with Spo2,ECG, Etco2, NIBP was connected.The child was induced by Sevoflurane priming at 8% and Oxygen at 4 litres. The child was then inserted with a size 1 LMA. Under strict aseptic protocols the child's right internal jugular vein was secured with a 5.5 f 2 lumen central line and during the procedure the infant's hemodynamics was stable through out and was brought out from sedation safely and shifted back to pediatric intensive care unit as per the Figures (A,B and C).



Lab test has shown:

Liver function test had the following results:

Total serum bilirubin 1.2 mg/dL;

Indirect serum bilirubin 0.8 mg/dL (direct),

0.4 mg/dL;

(SGPT) 28 U/L

(SGOT) 64U/L.

## **Discussion**

The combination of thrombocytopenia and absent radii was first described by Greenwald and Sherman in 1929 and as a syndrome by Hall et al in 1969.

The most common features in a tar syndrome are thrombocytopenia which presents in almost 100% of patients and becomes symptomatic mostly in the first few months of life. Platelets usually range between 15000-30000 in infancy and mostly improve in due course of age. The impaired bone marrow production of platelets in spite of normal thrombopoietin production makes way for thrombocytopenia. The main cause of death in a neonate is usually haemorrhage.

The absence of unilateral or bilateral radius is another common manifestation (100%) in a Tar syndrome patient. The Lower limb anomalies also presents such as hip dislocations and ankylosis and foot deformities. Cardiac anomalies such as Tetralogy of fallot, VSD, ASD also makes a greater obstacle to tackle. Presence of cardiac diseases dictates specific technique incorporation and high risk for paradoxical embolism also has to be considered for this subset of patients.

Tar syndrome infants usually would have multiple failed venous cannulations in the ward and it necessitated the requirement of a central line in our case.

The infants would usually also require frequent blood sampling along with the administration of blood and blood products due to their thrombocytopenia. Due to the presence of underlying cardiac anomalies there is also high risk of paradoxical embolism.

In our case it was an emergency requirement of invasive lines as severe dehydration has further complicated and an evaluation of multiorgans was quite not possible. Ultrasound guidance provided a safe ground for attaining the central line.

## **Conclusion:**

Good organization, communication, updated knowledge on rare diseases and teamwork of anaesthesiologists with pediatricians and pediatric genetics department including careful planning are the cornerstones of safe management of a child with Covid 19 and a rare disorder.