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Anaesthesia Management of Shwachman - Diamond Syndrome Child Posted for Dental Rehabilitation

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Abstract: ***Shwachman - Diamond syndrome (SDS) is an autosomal recessive disease which results in inherited bone marrow failure (IBMF) and is characterized by pancreatic dysfunction, skeletal abnormalities. and diverse clinical phenotypes. In the present study, we reviewed the internationally published reports on SDS patient, in order to summarize the clinical features, epidemiology, and intraoperative anaesthesia management of SDS. It is the most common cause of pancreatic insufficiency in children after cystic fibrosis. We report a child with classical SDS who was posted for Complete Dental Rehabilitation***

Keywords: Shwachman syndrome, Lipomatosis, Pancreatic insufficiency

1. Case Report

A 3 - year - old boy presented to us with dental caries. He also had frequent episodes of fever requiring antibiotics, recurrent oral ulcers, pruritic papular skin rash and dental caries. The child was a product of non - consanguineous marriage with normal birth weight and an apparently healthy elder sister. There was no family history of pancreatic insufficiency or hematological disorders. His physical examination revealed growth retardation (weight was constant 10.4 kg to 11kg on creon 10000 Units 3 times a day. {pancreatin 10000U cap}. On multivitamin and multi mineral syrup, 10mg vitamin k tablet**,** Vitamin E and vitamin C capsule. History of Stools 1 to 2 times /day, steatorrhea, mild pruritic and dental caries

A diagnosis of Shwachman - Diamond syndrome was made and he was started on pancreatic enzyme and fat soluble vitamin supplementations. On follow - up, steatorrhea settled and growth improved (1.5 kg increase in weight and 4 cm increase in height after 6 months). Intermittent neutropenia continued to occur on follow - up but no evidence of serious infection SDS is a rare autosomal recessive disorder characterized by pancreatic insufficiency, hematologic dysfunction, and skeletal abnormalities. Neutropenia is present in 98% of SDS patients, leading to frequent infections. Other organ systems are commonly involved, presenting a challenge in the perioperative period. Patients with SDS often require multiple surgical interventions, and chronic pain states are common, making the achievement of adequate pain control an ongoing concern. He was scheduled for a Complete Dental Rehabilitation, preoperatively, the patient was assessed. Paediatrician fitness taken for surgery and anaesthesia. Lab reports noted normal except neutropenia.

Plan of Anaesthesia:
Induction of anaesthesia was accomplished with propofol 40mg and Atracurium 8mg. Inj Fentanyl 20mcg was used as analgesia After the airway was secured with nasal endotracheal tube 4, 5 cuffed, sevoflurane and dexmedetomidine were used for maintenance of anesthesia. At the end of the procedures, the patient was extubated and transported to the recovery room. Postoperatively, the patient was controlled by diclofenac suppository 12, 5mg for pain control. He was discharged home next day

Patients with SDS often have recurrent respiratory infections, decreased FRC. Respiratory collapse can occur if caution is not used when administering opioids

For these reasons, a multi - modal approach to perioperative pain control in these challenging patients is suggested.

2. Discussion

When a patient presents with steatorrhea with recurrent respiratory symptoms, the diagnosis first considered is cystic fibrosis. As infections in our patient were not restricted to lungs, the possibility of immune deficiency was also considered. However, steatorrhea with intermittent neutropenia suggests the diagnosis of SDS. Patients with SDS experience recurrent viral, bacterial and fungal infections, including otitis media, bronchopneumonia, osteomyelitis, skin infection and septicemia. The quantitative and qualitative defects in neutrophils contribute to these infections in SDS [6, 7]. Failure to thrive is a common manifestation because of malabsorption, recurrent infections and metaphyseal dystoses. The diagnostic criteria of SDS, as laid down by Dror and Freedman, require documentation of exocrine pancreatic dysfunction and characteristic hematological abnormalities [8]. Although not done in our case, genetic analysis can be performed for confirmation. Shwachman - Diamond syndrome (SBDS) gene is located at chromosome 7q11 [6, 9]. Ultrasonography shows normal sized pancreas with increased echogenicity of the silhouette but CT and MRI reveal lipomatosis of the pancreas with greater accuracy [6]. For gastrointestinal manifestations, the mainstay of treatment is pancreatic enzyme therapy, medium - chain triglyceride, and fat soluble vitamin supplements. With this treatment, steatorrhea resolves and body weight increases but growth is not generally accelerated [6]. Exocrine pancreatic function tends to improve with time in almost half of the patients, but hematological problems deteriorate [6]. Our patient showed

response to enzyme supplements but continued to have problems related to neutropenia. For the treatment of hematological abnormalities in SDS, stem cell transplantation [10] and bone marrow transplantation have been reported [3]. The projected median survival of patients with SDS is 35 years and the main reason of untimely death is hematological (bone - marrow failure, myelodysplastic syndrome and acute myeloid leukemia).

3. Conclusion

The diagnosis of Shwachman - Diamond syndrome (SDS) should be strongly considered in any child who presents with steatorrhea, growth retardation, and intermittent or persistent neutropenia

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