

A CASE REPORT: PIERRE ROBIN SYNDROME**Sonia Kumari Singh^{1*}, Abijith Rajendra Babu¹, Britto DuraiSingh² and Sheik Haja Sherief³**¹Pharm D. Interns, Department of Pharmacy Practice Nandha College of Pharmacy, Perundurai Main Road, Erode, Tamil Nadu.²Clinical Pharmacist, GMCH hospital, Coimbatore.³Department of Pharmacy Practice, Nandha College of Pharmacy, Perundurai Main Road, Erode, Tamil Nadu.***Corresponding Author: Dr. Sonia Kumari Singh**

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Article Received on 16/09/2019

Article Revised on 06/10/2019

Article Accepted on 27/10/2019

ABSTRACT

Pierre Robin Syndrome is characterized by triad of micrognathia, glossoptosis and U- shaped cleft palate. We report a case of 3month baby who referred to our tertiary care hospital with a diagnosis of pierre robin syndrome and was further managed with surgical procedure.

KEYWORDS: cleft palate, micrognathia, glossoptosis, pierre robin syndrome(PRS).**INTRODUCTION**

Pierre Robins syndrome has three main features of micrognathia, macroglossia and cleft palate first described by Pierre Robin a French dental surgeon in 1923^[1] PRS often develops upper airway obstruction or feeding difficulty secondary to micrognathia, glossoptosis, or a shifted tongue that comes in contact with the pharyngeal wall.^[2] Usually, progressive airway obstruction might become more noticeable in the second month of life. This "sequence of events" is the reason why the condition has been classified as a sequence.^[4,5,6] The exact causes of Pierre Robin sequence are unknown.^[1] Possible mechanisms for the sequence include genetic causes; low volume of amniotic fluid (oligohydramnios), which may limit chin growth; weakness of the facial muscles (myotonia); or connective tissue disease.^[3] The management for PRS is either conservative treatment or surgical interventions.

CASE PRESENTATION

A 3-months-old 4.5kg female baby was referred to our tertiary care hospital with working diagnosis of pierre robin sequence. Baby was delivered by LCSC in outside hospital. Upon admission baby had complaints of retrognathia, cleft palate, glossoptosis along with breathing and feeding difficulty. Before 2 months patient had a history of tongue tie suture done. Baby had strider, difficulty in breathing and feeding for which glossopexy was done under general anaesthesia. After glossopexy baby had history of mechanical ventilation for 3 days and extubated. Followed by which baby developed desaturation and bradycardia. Baby was shifted with medical support from his native hospital. Now admitted here for definitive management. During this period baby

being nursed in prone position to avoid respiratory distress. During transfer baby shows decrease oxygen saturation which settled with 4 L oxygen. Baby also shown a sign of respiratory infection for which she was started on inj meropenem by transferring team.

Day 1 patient findings were, PO₂ was 95-100% in prone position. Lateral examination shows micrognathia, retrognathic, mandible, cleft of secondary palate present, glossoptosis. Clinical and biochemistry report shows CRP was high 84.07mg/L, RBC 3.1x10¹²/L, Hb 9.2g/dl, PCV 27.6%, MCV 89 Fl, MCH 29.6 pg, MCHC 33.2%, PLT 519x10⁹, WBC 9.7 x10³/L, neutrophils 49.3%, lymphocytes 38.5%, monocytes 7.1%, eosinophil 4.1%, basophil 1.0% while blood culture shows no growth and advised for ABG analysis.

Day 2, ABG analysis done shows normal result. Patient was continued on meropenem 60mg tid for 5days. Advised for RFT which shows slight increase in potassium level 5.62mmol/L. Decided for surgical procedure next day advised to prepare baby for procedure.

Day 6, preoperative medications, amikacin 23mg and paracetamol suppository 80mg were given. Osteotomy done by using oscillating saw. Procedure was uneventful. After surgical procedure patient kept under conservative care and regular observation was given.

Day 7, CBC was repeated, report show decline in hemoglobin count 6.9g/dl. Patient was given 10 unit of PRC transfusion. Patient was continuously followup for 5 days. Baby shown prominent improvement. On 12th

day patient was discharged without any specific complaints.

DISCUSSION

Pierre robin sequence also termed as Pierre robin syndrome (PRS). PRS refers to the association of micrognathia and glossoptosis and is characterized by varying degree of upper airway obstruction. Usually airways obstruction or breathing difficulty in 2nd month of life.

PRS may occur alone or in association with other syndrome such as stickler syndrome. The cause of PRS may be genetically isolated, either recessive or dominant autosomal condition. In our case no association with genetic condition were observed. PRS can be life threatening during neonatal period with onset of airway obstruction, which can occur anytime right after birth. If left untreated may leads to respiratory tract infection, chronic hypoxia, cyanosis, apnea episodes and feeding difficulty. Complications of PRS are cerebral hypoxia, corpulmonale and right heart failure.^[5,6]

Most of the patients with PRS get relieved by conservative measures however patient with pronounced micrognathia, retrognathic mandible, U shaped cleft palate and prolonged endotracheal intubation needs surgical correction or else in less severe cases conservative treatment is sufficient. As in our case surgical intervention were done to manage the complaints of neonates.^[8,10]

CONCLUSION

All the case of PRS should be thoroughly investigated to differentiate and diagnose in association with other syndrome and to formulate the further line of management.

ABBREVIATION: PRS- Pierre robin syndrome.

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