

## CASE REPORT

# A Newborn With Constriction Ring Syndrome: A Case Report

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### ABSTRACT

We report a rare case of constriction ring syndrome in a 3515 grams full term female newborn who was born with a constriction ring encircling the fingers of her right hand diagnosed as acrosyndactyly. It is a rare congenital malformation and the risk factors are still unknown. The neonate had severe swelling of her third and fifth fingers, and skin breaches. There were intrauterine amputations in her second and fourth fingers. The X-ray images showed deformities in both the size and shape of the bones of her fingers. No additional abnormalities or comorbidities were found. The neonate was referred to a better equipped hospital because of impending necrosis from autoamputation. During anamnesis, the mother recalled that she took haloperidol and trihexyphenidyl until 15 weeks' gestation. There was neither history of fever, trauma, intake of herbal preparations, nor family history of deformity. Etiology and risk factors of congenital constriction ring syndrome are still under discussion. Early intervention is recommended for a successful outcome.

**Keywords:** Congenital constriction ring, Amniotic band syndrome, ADAM Syndrome, Autoamputation, Early intervention

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### INTRODUCTION

Constriction ring syndrome (CRS) is an uncommon congenital anomaly with multiple disfiguring and disabling manifestations (1). CRS, also known as ADAM complex, Streeter dysplasia, constriction band, and amniotic band syndrome is anomaly involving constricting bands that encircle, either partially or completely a digit or extremity (1,2). Patients present with a wide spectrum of deformities including simple ring constrictions, minor digital defects, and occasional craniofacial and visceral defects. It occurs in approximately 1 in 1,200 to 1 in 15,000 live births. Incidence of other anomalies and comorbidities, for example, club feet or cleft lip and palate have been reported, although no direct genetic etiology has been established (3).

### CASE REPORT

We report a rare case of congenital ring syndrome in a 3515 grams full term female newborn who was born with a constriction ring encircling the fingers of her right hand diagnosed as acrosyndactyly. Her mother was 37 years old gravid 4 para 3 with low social economy.

Fibrous bands were present within the ring. The bands measured 0.1-0.5 cm in width (Fig. 1). Granulation tissue was present in the depths of the ring. The neonate had severe swelling of her third and fifth fingers, and skin breaches. There were intrauterine amputations in her second and fourth fingers (Fig. 2). The X-ray images (Fig. 3) showed narrowing of soft tissue on the proximal right fingers causing enlargement of the distal digits. There



**Figure 1: Constriction ring with encircling fibrous band**



**Figure 2: Intrauterine amputation of finger**



**Figure 3: X-ray of right (CRS) and left hand (normal)**

was deformity in both the size and shape of several bones of her fingers. There was some indentation in her right arm. There were no signs of acute limb ischemia at birth. The baby has no additional abnormalities or comorbidities. There was no family history of similar anomalies. The neonate was referred to Orthopedic and Traumatology Clinic, Sardjito General Hospital as a better equipped hospital because of impending necrosis due to autoamputation. Z-plasty was done to release contractures (Fig. 4). Reconstruction and separation of the finger will be held six months later.

The prenatal history was remarkable because the mother took haloperidol 0.5 mg/12 hours and trihexyphenidyl 5 mg/12 hours for 9 years, from previous delivery until 15 weeks' gestation of this pregnancy. She took them because diagnosed with post-partum depression



**Figure 4: Post-Z-plasty of right hand**

(F53.0). She had implant as contraception for 13 years between her previous pregnancies and had removed 3 years before this pregnancy. The mother did routine antenatal care with a midwife. Ultrasound was done once at 22 weeks' gestation. Ultrasound was done by obstetrician without any further result that informed to mother. No genetic testing was done. There was neither history of fever, trauma, bleeding, intake of herbal preparations, nor family history of hand and foot deformity.

The labor was pervaginam without any complications. She was born with Apgar score 7/8/9 and mild asphyxia. A 3-D digital image replication of her hand will allow a retrofit prosthetic which can be enlarged occasionally as she grows reducing any stigma in the future from her disability.

## DISCUSSION

The prevalence of the syndrome varies in different studies. It was once reported to have an incidence of 1:100,000, while recent literature indicates the incidence today as 1:1,200 - 1:1,500 births. The observed geographic difference in birth prevalence could be a useful indication to study specific genetic and environmental candidate factors that may pollute specific populations (1).

No sex predilection has been determined. Prenatal risk factors associated with amniotic band syndrome include prematurity (<37weeks), low birth weight (<2500g), maternal illness in pregnancy or placental hemorrhage in trauma, and attempted abortion in the first trimester are highly associated finding (1). No genetic predisposition has been found and occurrences of ring syndrome have

not been seen across generations.

There are many previous studies of adverse events associated with the use of haloperidol during pregnancy like microphthalmia, trisomy 13 and 18, gastrochisis, renal dysplasia, dyskinesia in the newborn, neonatal hypothermia, acute dystonia, withdrawal syndrome, aortic valve defect, and isolated limb defect (2–5). But, they were also exposed to other drugs and there is no clear causal relationship with haloperidol. A case control study conducted by European Network of teratology Information Services showed that the rates of major malformations did not differ between the control and haloperidol groups (3). A review of antipsychotic during pregnancy concluded that there was still not enough data to determine whether it is safe or not (4). There are not enough data on trihexyphenidyl in human pregnancy or animal. There were minor malformations found in a study of anticholinergic exposure during first trimester but only nine of 2,323 patients that took trihexyphenidyl. A case report of using haloperidol and trihexyphenidyl during three consecutive pregnancies showed no event at all in babies (2).

A wide variety of clinical presentations have been reported contributing to a number of differential diagnoses. The incidence of malformation seen in the hand is twice more common accompanied by foot deformities. The difference in deformities appears to be related to time and extent of injury as well as location of the constriction rings (5).

Ultrasound has been used to diagnose CRS prenatally, but the phantom calibration for digital X-ray tomosynthesis can be challenging. Early diagnosis at 12 gestational weeks can be confirmed by additional examinations. By the 3rd month of gestation, most defects appear in routine ultrasound examinations (5). Terminal syndactyly and autoamputation are important ultrasound diagnostic criteria.

Surgery is the mainstay of treatment. Release of the constriction rings and acrosyndactyly can improve prognosis and restore basic hand functions such as grip and pinch. Z-plasty can be conducted for more severe constrictions and distal deformity while treatment in mild cases of CRS may not require treatment (1). In our case, the early intervention was successful and prognosis excellent.

## CONCLUSION

The etiology and risk factors of congenital constriction ring syndrome are still under discussion. Early intervention is desirable for a successful outcome.

## ACKNOWLEDGEMENT

We would like to thank Meirizal Hasan, MD for his postsurgery photo.

## REFERENCES

1. Kawawura K, Chung KC. Constriction band syndrome. *Hand Clin.* 2009;25:257–64.
2. Mendhekar DN, Andrade C. Uneventful use of haloperidol and trihexyphenidyl during three consecutive pregnancies. *Arch Womens Ment Heal.* 2011;14:83–4.
3. Diav-citrin O, Shechtman S, Ornov S. Safety of haloperidol and penfluridol in pregnancy: a multicentre, prospective, controlled study. *J Clin Psychiatry.* 2005;66:317–22.
4. Odhejo YI, Jafri A, Mekala HM, Hassan M, Khan AM, Dar SK, et al. Safety and Efficacy of Antipsychotics in Pregnancy and Lactation Alcoholism & Drug Dependence. *J Alcohol Drug Depend.* 2017;5(3):267–83.
5. Wahegaonkar C, Patil B, Dogra BB, Agarkhedkar N. Presentation and treatment of congenital constriction ring syndrome: case series of 12 patients. *Int J Res Med Sci.* 2016;4:2181–4.