Introduction
Congenital nasal pyriform aperture stenosis (CNPS) is a rare, potentially lethal form of neonatal nasal obstruction. It arises in the fourth month of fetal development because of an overgrowth of the nasal process of the maxilla and may present as an isolated malformation or may be associated with other craniofacial anomalies. CNPS manifests as nonspecific symptoms of nasal airway obstruction, such as apnoeic crisis, episodic cyanosis, and inability to nurse. Conservative treatment usually fails. The most effective treatment for these patients is a surgical correction of the stenosis through endoscopic endonasal or endooral sublabial approach followed by a nasal stenting.

Case report
We report a case of a male infant born at 39 weeks by acute section because of imminent fetal hypoxia. The infant early showed respiratory distress that was presented with mild inspiratory stridor. ENT examination excluded choanal atresia or other abnormality of upper aerodigestive tract. The only pathology was oedematous nasal mucosa in both sides of nasal cavity. The conservative therapy failed. The next step in diagnostic procedure was CT and MRI scan because of suspicion of CNPS. At 32 days after birth we performed endo-nasal pyriform stenosis enlargement using gum elastic bougies for dilatation up to size 19Ch. In second part of surgery the PVC nasal stents (o.d. 5.2 mm, i.d. 3.5 mm, length 70 mm) were sutured to the columella to maintain airway patency and to stabilize the surgical results. The nasal stents were removed on seventh postoperative day. The phenotype of newborn didn’t reveal any genetic stigmatisation, no other associated congenital abnormality was found. The patient was discharged with no complication at age 43 days.

Conclusions
The congenital nasal pyriform stenosis can be one of the causes of inspiratory dyspnea and respiratory distress in early postnatal life. Although this anomaly is rare, early diagnosis and therapy is mandatory because it’s potentially life – threatening condition.

References