

CASE REPORT

Complex Airway Management in Pierre Robin Sequence with Failed Intubation: Case Report and Lesson Learned

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ABSTRACT

Background : Pierre Robin Sequence (PRS) is a rare condition with a global prevalence of 1 in 8,500-14,000 live births, characterized by micrognathia, glossoptosis, and upper airway obstruction. Reporting this case is essential to enriching the literature on PRS management, especially in Indonesia, where case data is still limited.

Case Illustration : A 1 year ten months old child with PRS, palatoschizis, laryngomalacia, secondary hypothyroidism, and mandibular hypoplasia. The patient will undergo mandibular osteodistraktion to address airway obstruction and improve facial structure. Since birth, the patient has shown feeding difficulties and airway obstruction. At five months of age, an echocardiography examination revealed a patent ductus arteriosus (PDA). The patient was fasted for 6 hours before surgery, intravenous access was established, and premedication with midazolam 0.05mg/kgBB. Preoxygenation, induction with ketamine 1mg/kgBB, intubation with spiral ETT No. 3.5 and atracurium 0,5mg/kgBB were given, and maintenance anaesthesia using sevoflurane. Close monitoring was done to ensure the patient's stability. Postoperatively, the patient was transferred to the intensive care unit with intravenous paracetamol analgesic and close monitoring to prevent complications.

Conclusion This case highlights the importance of meticulous airway planning in children with Pierre Robin Sequence (PRS) and a history of failed intubation. Successful anaesthetic management was achieved through preoperative airway assessment, adequate preoxygenation, successful tracheal intubation before administration of neuromuscular blockade, and maintenance with sevoflurane. Given the potential for difficult ventilation and intubation in PRS, appropriate preparation for advanced airway techniques and multidisciplinary collaboration are essential.

Keywords: Anesthesia management; Mandibular osteodistraktion; Palatoschizis; Pierre Robin Sequence

INTRODUCTION

Pierre Robin Sequence (PRS) is a clinical condition characterized by micrognathia (mandibular hypoplasia), glossoptosis (downward displacement of the tongue), and upper airway obstruction¹⁻³. The condition causes impaired mandibular development, resulting in the tongue falling into the throat and blocking the upper airway. PRS is often associated with cleft palate (palatoschisis), caused by mandibular hypoplasia early in fetal development, inhibiting palatal closure at the midline. PRS was first described in 1891, and Pierre Robin published the first case in 1923. With a global prevalence of approximately 1 in 14,000 babies, no data on PRS in Indonesia exists. PRS is often associated with other syndromes, such as fetal alcohol syndrome, Stickler syndrome, velocardiofacial syndrome, and Treacher-Collins syndrome^{1,4,5}.

In neonates with PRS, respiratory distress often presents as stridor, retractions and cyanosis, as well as feeding difficulties, gastroesophageal reflux, aspiration and failure to thrive. While pronation or lateral positioning can help reduce airway obstruction in about 70% of cases, some require further

measures, such as nasopharyngeal or gastrostomy tube insertion and emergency tracheostomy in severe acute airway obstruction. Mandibular osteodistraction, a technique introduced by McCarthy in 1989, has become the primary option to treat PRS by improving airway obstruction and facial cosmetics and correcting malocclusion through jaw elongation and tongue displacement^{1,4,5}.

Anaesthesia management in PRS patients presents complex challenges and requires careful preparation and effective team coordination. Therefore, this case report aims to highlight the challenges of anaesthetic management in PRS patients undergoing mandibular osteodistraction, focusing on optimal anaesthetic strategies and techniques to ensure the safety and success of the procedure.

CASE ILLUSTRATION

A child aged 1 year and 10 months with a diagnosis of Pierre Robin Sequence (PRS) with palatoschisis, laryngomalacia, secondary hypothyroidism and mandibular hypoplasia. The patient will undergo mandibular osteodistraction. At birth, there was an anatomical abnormality of the palate and difficulty drinking. The patient was then referred to RSCM and diagnosed with Pierre Robin Sequence. At 5 months,

the patient underwent an echocardiography examination and found a patent ductus arteriosus (PDA). In addition, an endocrine examination was performed and diagnosed with secondary hypothyroidism. The patient was planned for palatoplasty at RSCM, but it failed because intubation was not possible. The patient was born to a mother (33 years old) at 37 weeks gestation. At birth, the patient cried immediately. V (-) A (-) C (+) T (-) E (-) R (-) L: PDA. There was no history of drug allergy (-). The patient was taking Eutirax at a dose of 1x25 mcg). Previous medical history included Pierre Robin Sequence, palatoschizis, laryngomalacia, secondary hypothyroidism, and mandibular hypoplasia.

On physical examination, the patient weighed 6.7 kg and had a GCS E4M6V5 level of consciousness. Pulse rate was 126 beats per minute, respiratory rate was 28 beats per minute, body temperature was 36.4°C, and oxygen saturation (SpO₂) was 99% with free air. General examination showed non-anemic conjunctiva and noncrystallized sclera. There was a cleft palate, and an NGT was in place. On the thorax, chest wall movement was symmetrical, with good vesicular breath sounds (VBS) on both sides without rhonchi or wheezing. S1 and

S2 heart sounds were regular with no murmurs. The abdomen is flat and soft, and bowel movements (BU) are standard. Extremities were warm, with a capillary filling time of less than 2 seconds. The patient's COPUR index was 11.

The patient's laboratory examination showed a haemoglobin (Hb) level of 12.2 g/dL and hematocrit (Ht) of 38.4%. The leukocyte (L) count was 9,590 cells/ μ L and platelets (Tr) 389,000 cells/ μ L. Prothrombin time (PT) was 12.6 seconds with an international normalized ratio (INR) of 0.88 and activated partial thromboplastin time (aPTT) of 24 seconds. Blood urea level was 22.1 mg/dL, and creatinine was 0.51 mg/dL. Liver enzymes showed SGOT 40 U/L and SGPT 23 U/L. Electrolyte sodium (Na) levels were 139 mEq/L, potassium (K) 4.2 mEq/L. Thyroid hormones recorded T3 of 1.7 ng/mL, fT4 of 1.2 ng/dL, and TSHs of 0.6 μ IU/mL.

Chest X-ray examination showed that the corpus and pulmo were within normal limits. Rhinolaryngo fibre optic (RLFO) examination revealed laryngomalacia and cleft palate. Echocardiography showed atrial situs solitus, normal pulmonary and systemic venous drainage and four balanced heart chambers. There was patent foramen ovale (PFO) and trivial tricuspid regurgitation

(TR), but no mitral regurgitation (MR), intact interventricular septum (IVS), no aortic regurgitation (AR), pulmonic regurgitation (PR), or patent ductus arteriosus (PDA). The left aortic arch was normal without coarctation of the aorta, with well-contracted ventricles, no paradoxical motion, and no pericardial effusion. The conclusion of this examination was the spontaneous closure of the PDA.

Based on these examination results, the patient was diagnosed with Pierre Robin Sequence with palatoschizis, laryngomalacia, secondary hypothyroidism, and mandibular hypoplasia. ASA III. Based on the preoperative evaluation, the patient was considered to have an anticipated difficult airway due to severe mandibular hypoplasia associated with Pierre Robin Sequence, palatoschizis, laryngomalacia, and a previous history of failed tracheal intubation during a planned palatoplasty. In this case, difficult airway was determined based on the patient's clinical and anatomical characteristics and previous airway management history. No specific difficult-airway scoring system was used. Therefore, airway management was planned with preparation for potential difficulty in both ventilation and

tracheal intubation as well as potential problems such as desaturation, laryngeal/bronchial spasm, hypothermia, hypoglycemia, and bradycardia. In the preoperative phase, patients were fasted for 6 hours before surgery and given intravenous access. Premedication was performed by administering 0.05 mg/kgBB (0.3 mg) of Midazolam. Monitoring included blood pressure, precordial, IBP, temperature, saturation, urine output, EtCO₂, GDS, and crystalloid fluid preparation at a rate of 28 cc/hour.

In the intraoperative phase, preoxygenation was performed with a 50:50 mixture of oxygen and nitrous oxide. Sevoflurane was administered gradually until the patient lost consciousness. A face mask was applied in a reversed position to accommodate the patient's facial anatomy, while bag-mask ventilation was performed. During induction, the patient experienced a period of oxygen desaturation. A modified nasopharyngeal airway (NPA), prepared from a previously measured and appropriately shortened endotracheal tube, was subsequently inserted to facilitate airway patency and ventilation. Ketamine 7 mg was then administered.

Tracheal intubation was performed using a flexible fiberoptic

bronchoscope. The trachea was visualized, with the glottic/tracheal axis noted to be markedly anterior, which contributed to the anticipated difficulty in advancing the endotracheal tube. A size 3.5 spiral endotracheal tube was subsequently advanced under fiberoptic guidance. After successful placement of the ETT was confirmed, atracurium 3.5 mg was administered. Maintenance anaesthesia was performed with 2-3 vol% sevoflurane in a 50:50 O₂ mixture. Monitoring during surgery included O₂ saturation, NIBP, EtCO₂, urine output, GDS, and ETT retention. The patient was transferred to the intensive care unit in the postoperative phase. Analgesia was given with 4x200 mg paracetamol intravenously.

DISCUSSION

Pierre Robin Sequence (PRS) is characterized by a clinical triad of micrognathia (mandibular hypoplasia), glossoptosis (downward displacement of the tongue), and upper airway obstruction¹⁻³. Clinically, this results in a small and underdeveloped mandible, causing the base of the tongue to fall back into the throat, eventually leading to upper airway obstruction. Globally, PRS is estimated to occur in 1 out of 8,500-

14,000 newborns annually^{1,5}. There is no data on PRS in Indonesia.

The aetiology of PRS is usually classified into isolated and syndromic PRS. Non-syndromic PRS is associated with chromosome 2, 4, 11, or 17 mutations. Some evidence suggests that mutations in SOX9 or KCNJ2 (on chromosome 17) can affect the development of facial structures and cartilage development, leading to PRS. Cleft palate is associated with deletions in 2q and 4p, and duplications in 3p, 3q, 7q, 8q, 10p, 14q, 16p, and 22q. Micrognathia is associated with deletions in 4p, 4q, 6q, and 11q and duplications in 10q and 18q. Syndromic PRS accounts for 60% of PRS cases. There are 34 syndromes associated with syndromic PRS, the most common being Stickler syndrome¹.

Pierre Robin Sequence (previously known as Pierre Robin Syndrome) is now accurately termed a sequence because it involves one initial malformation that leads to a chain of subsequent events causing other anomalies. In contrast, a syndrome is a set of anomalies that arise independently due to a single underlying pathogenesis. In PRS, micrognathia is the initial abnormality that causes glossoptosis, airway obstruction and cleft palate¹.

There are several common theories behind the development of hypoplastic

mandible. The Mechanical Theory states that the mandible typically grows ventrally and inferiorly in early pregnancy, around the seventh week of gestation. If the mandibular growth is abnormal, the tongue cannot follow the typical growth trajectory and obstruct the closure of the palatal cleft by the 11th week of pregnancy. This leads to glossoptosis, which eventually results in upper airway obstruction. The Neurological Maturation Theory proposes a delay in the neuromuscular development of the tongue, which in turn does not stimulate the mandible to grow correctly or the palatal shelves to fuse. Furthermore, the Mandibular Compression Theory suggests that external forces cause the fetal head to flex, pressing the mandible against the chest and preventing proper growth. This may be due to multifetal pregnancy, uterine abnormalities, or oligohydramnios. The tongue usually grows and shifts backwards, potentially causing upper airway obstruction. The tongue tip may also obstruct palatal fusion, creating a cleft palate¹.

Pierre Robin Sequence (PRS) poses many challenges for anesthesiologists. In patients with PRS, may present with difficult mask ventilation and tracheal intubation

because of their craniofacial anatomy. Maintaining a ventilatory seal using a face mask can also be challenging due to facial deformities. Direct laryngoscopy and tracheal intubation are difficult in infancy but generally become more manageable with age and mandibular growth⁵.

Postoperatively, PRS patients may experience spontaneous airway collapse. This can occur due to airway obstruction, potential obstructive sleep apnea (OSA), chronic hypoxia, and increased opioid sensitivity. Paradoxical breathing patterns, intercostal retraction, subcostal recession, sternum retraction, and tracheal tugging can diagnose this. Neonates with PRS are more sensitive to opioids. Infants suffering from OSA due to glossoptosis have increased opioid sensitivity due to the upregulation of opioid receptors in the brainstem. Patients with severe obstruction have significantly lower opioid requirements, and careful dose adjustment is necessary⁵.

Feeding difficulties, swallowing disorders, and gastroesophageal reflux are often complicated by bronchial microaspiration and lung infections. Therefore, aspiration precautions should be implemented before elective procedures. PRS patients generally experience malnutrition and have a high frequency of

failure to thrive due to feeding and airway difficulties⁵.

Early management and thorough evaluation are crucial in Pierre Robin Sequence (PRS) patients. PRS patients are typically admitted to the neonatal intensive care unit (NICU) for further assessment and management (Figure 2). This process requires a multidisciplinary approach involving various specialists, including neonatology, craniofacial/plastics, ENT, genetics, nutrition, speech pathology, and occupational therapy. Flexible nasolaryngoscopy is often performed at the bedside to evaluate the degree of airway obstruction, while microlaryngoscopy or bronchoscopy in the operating room may also be necessary^{6,8}.

The primary priority in out^{6,7}. management is to relieve airway

obstruction. Various options can be considered, such as placing the patient in the prone position to allow the mandible and tongue to fall forward, thereby reducing obstruction. Other alternatives include using a nasopharyngeal airway (NPA) or the distal tip of a modified endotracheal tube placed intranasally and in the distal oropharynx. In more severe cases, endotracheal intubation or tracheostomy may be required. Fiberoptic endoscopic evaluation of swallowing (FEES) is usually performed to assess swallowing ability, and if necessary, supplemental feeding can be provided through a nasogastric tube (Figure 1). Gastroesophageal reflux therapy should be initiated, and associated anomalies or syndromes should be identified and ruled

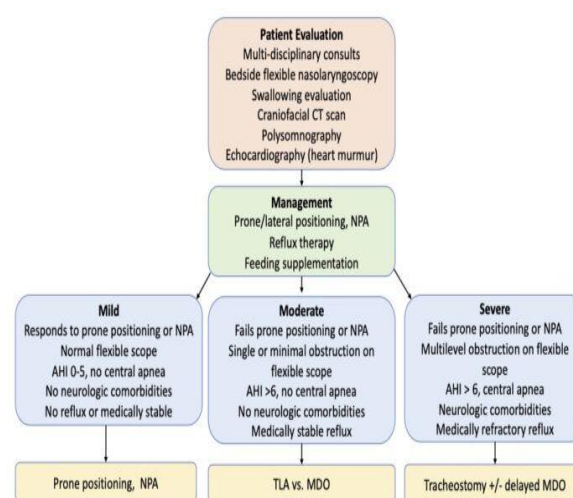


Figure 1. Flow of evaluation and management of patients with PRS⁶

NPA = nasopharyngeal airway, AHI = apnea-hypopnea index, TLA = tongue lip adhesion, MDO = mandibular distraction osteogenesis

In patients with Pierre Robin Sequence (PRS), airway obstruction can vary in severity, ranging from oropharyngeal obstruction caused by glossoptosis to hypopharyngeal obstruction due to the collapse of the epiglottis and the base of the tongue (EBT). Additionally, laryngomalacia, subglottic pathology, or tracheal issues can also occur. In nonsyndromic PRS patients, mandibular growth often catches up, and airway obstruction usually improves with age. However, this is only sometimes the case in syndromic PRS patients, where these problems may not resolve over time⁶.

In the preoperative evaluation of patients with Pierre Robin Sequence (PRS), gathering a detailed birth history, including APGAR scores and any episodes of apnea, cyanosis, or feeding difficulties during the early neonatal period, is essential. Physicians should also inquire about the need for prone positioning to manage airway obstruction. The physical examination should focus on the airway, including assessing the degree of micrognathia, cleft palate, glossoptosis, and airway obstruction in various positions. Additionally, heart murmurs and associated anomalies or syndromes should be ruled out. The results of flexible

nasolaryngoscopy, often performed at the bedside, should be reviewed to identify patent choanae, the degree of oropharyngeal obstruction, EBT collapse, and laryngeal abnormalities. Previous airway management techniques and diagnostic studies such as craniofacial CT, polysomnography, and echocardiography (if there is a heart murmur) should also be reviewed^{6,8}.

In preoperative preparation, the availability of equipment for complex airway management must be ensured, including various types of NPAs, supraglottic airways (SGAs), video laryngoscopes, and flexible bronchoscopes. The presence of an ENT surgeon and additional anesthesiologists at induction should be considered to ensure optimal management⁶.

During anaesthesia induction in PRS patients, various airway management techniques have proven effective, including minimal sedation, no sedation, or after induction of general anaesthesia. Maintaining spontaneous ventilation is crucial. Induction is typically done with volatile agents and intravenous propofol, with patients usually maintaining spontaneous ventilation until intravenous access is secured and the chosen airway technique is applied. Rigid

laryngobronchoscopy is performed while the patient is breathing, with anaesthesia maintained using volatile anaesthesia or intravenous propofol and short-acting opioids.

In this case, inhalational induction with sevoflurane was performed while attempting to maintain spontaneous ventilation. Because mask ventilation was difficult and transient oxygen desaturation occurred, a modified nasopharyngeal airway was inserted to improve airway patency and ventilation. Once adequate airway management was established, ketamine was administered and tracheal intubation was performed under flexible fiberoptic guidance.

This technique was selected because of the anticipated difficult airway associated with Pierre Robin Sequence, mandibular hypoplasia, laryngomalacia, and a previous history of failed intubation. Fiberoptic guidance allowed direct visualization of the airway and facilitated tracheal tube placement despite the markedly anterior position of the tracheal axis.

Local anaesthesia of the larynx can help reduce the risk of laryngospasm. If tracheal intubation is required,

neuromuscular blockade may be necessary after the airway has been successfully secured.

In this case, atracurium 3.5 mg was administered only after successful fiberoptic-guided tracheal intubation and confirmation of endotracheal tube placement. Atracurium was selected for neuromuscular blockade in this case and was not administered before securing the airway because of the anticipated difficult airway. This approach was consistent with the principle of maintaining spontaneous ventilation during airway management until the trachea was secured.

Rocuronium or vecuronium is preferred if sugammadex is available for emergency reversal. Simple analgesics can be used, with opioid choices depending on the extent of surgery and the risk of airway compromise and postoperative apnea. Alpha-2 agonists are often used as analgesics to reduce opioid use during the procedure. Neuromuscular block monitoring is essential to ensure complete reversal before awake extubation. The airway management algorithm for PRS patients is shown in Figure 3, and fiberoptic intubation through an SGA is illustrated in Figures 4^{6,7,9-11}.

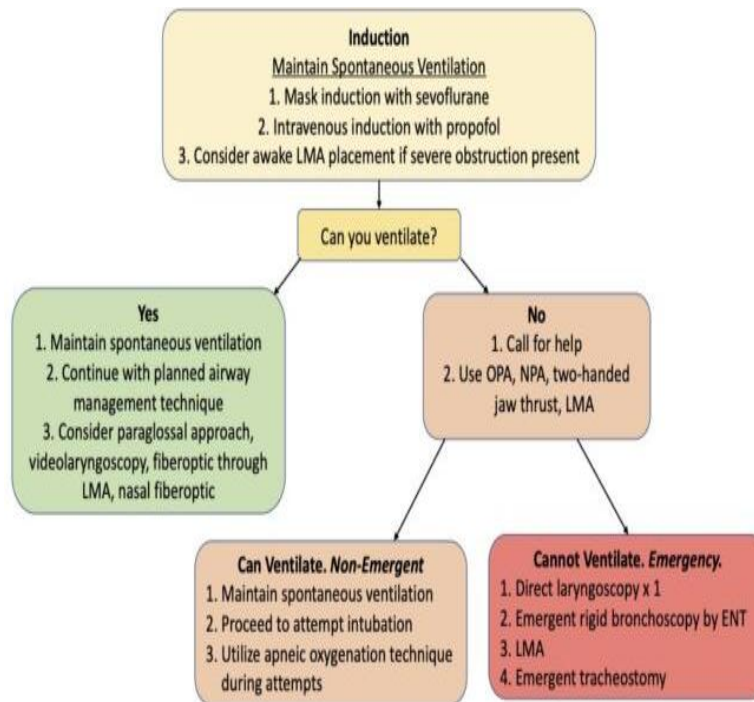


Figure 2 Algorithm for Airway Management in PRS Patients⁶
OPA = oropharyngeal airway, NPA = nasopharyngeal airway, LMA = Laryngeal Mask Airway

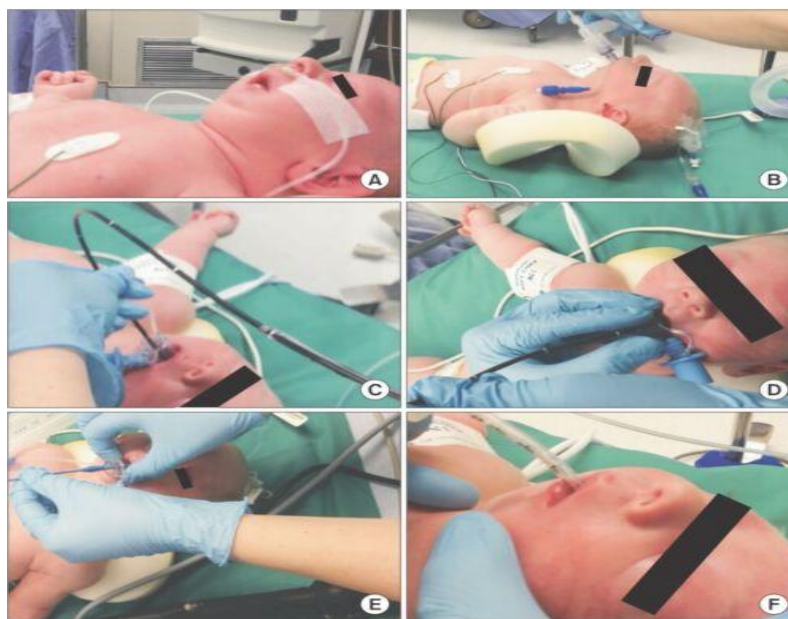


Figure 3 Steps for fiberoptic intubation through SGA in PRS infants⁶

(A) PRS baby before induction; (B) Laryngeal mask airway (LMA) Classic™ inserted while awake; (C) Flexible fiberoptic bronchoscope (FFB) placed through Air-Q® 1.0; (D) Endotracheal tube (ETT) inserted through the FFB with the patient under induction and relaxation; (E) Air-Q® Pusher used to remove the Air-Q®; (F) ETT in place.

During anaesthesia's maintenance and emergence stages, general anaesthesia is typically maintained using volatile anaesthetics, which may be supplemented with opioid infusions and alpha-2 agonists such as dexmedetomidine or ketamine. If tracheal extubation is planned, the patient must be fully awake before the procedure. Patients with Pierre Robin Sequence (PRS) undergoing procedures such as mandibular distraction osteogenesis (MDO) are usually intubated postoperatively for several days while the MDO device gradually functions^{6,11}.

In the postoperative phase, airway obstruction is a primary concern in managing PRS patients. These patients tend to be highly sensitive to opioids due to chronic hypoxemia, increasing the risk of postoperative respiratory complications. To reduce the risk of airway obstruction after extubation, a nasopharyngeal airway (NPA) should be considered. According to local practice, patients undergoing tongue-lip adhesion or mandibular distraction osteogenesis may require sedation in the neonatal intensive care unit (NICU). Patients needing non-invasive ventilation before surgery due to obstructive sleep apnea may require re-ventilation in the recovery area of the operating room. Additionally, apnea

monitoring should be available for patients who are opioid sensitive or have a history of obstructive or central apnea^{6,9}.

CONCLUSION

The anaesthetic management of this case, involving a child with Pierre Robin Sequence (PRS) with palatoschisis, laryngomalacia, secondary hypothyroidism, and mandibular hypoplasia, required thorough preoperative evaluation, careful intraoperative monitoring, and comprehensive postoperative care. Given the anticipated difficult airway and previous history of failed intubation, preparation for difficult mask ventilation and tracheal intubation was essential.

During induction, spontaneous ventilation was maintained, and transient oxygen desaturation was managed with a modified nasopharyngeal airway. Fiberoptic-guided tracheal intubation was subsequently performed successfully, and atracurium was administered only after confirmation of endotracheal tube placement. Close postoperative monitoring in the intensive care unit was maintained because of the risk of airway obstruction. This case highlights the importance of individualized airway planning, preparation for advanced airway techniques, and multidisciplinary

collaboration in the anaesthetic management of PRS.

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