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Intravenous Anesthesia in a Rare Pediatric Patient with Poland Syndrome: Case Report

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ABSTRACT

Poland syndrome (PS) is a very rare congenital anomaly characterized by hypoplasia or aplasia of the pectoral muscles and mammary hypoplasia and is associated with anomalies in the ipsilateral region of the body. Poland syndrome can increase the risk of developing intraoperative malignant hyperthermia; hence, special consideration is needed in the management of anesthesia in these patients. We report herein the management of general anesthesia for syndactyly release surgery in a child with Poland syndrome. A 1-year- and 11-month-old boy with Poland syndrome was scheduled to undergo manus dextra interphalangeal syndactyly release surgery under total intravenous general anesthesia using midazolam (0.1 mg/kg body weight), fentanyl (2 mcg/kg), and atracurium (0.5 mg/kg body weight) as induction agents, with maintenance using midazolam (0.3 mg/kg/h). Anesthesia can be performed at this age to prevent contractures and deformities and to avoid functional abnormalities. The anesthesia lasted 120 min with intraoperatively stable hemodynamics. After the operation, the patient was treated in a regular ward and discharged after 2 days of treatment. The use of total intravenous anesthesia with midazolam in children with Poland syndrome for the surgical release of manus dextra interphalangeal syndactyly is a good option and is also clinically beneficial.

KEYWORDS *pediatric anesthesia, Poland syndrome, intravenous anesthesia, malignant hyperthermia, general anesthesia.*

INTRODUCTION

Poland syndrome (PS) is a rare congenital anomaly with an estimated incidence of one in 30,000 to one in 80,000 live births, with the majority occurring in men and generally occurring on the right side of the chest compared to the left. Until now, there is no exact data on the cases of Poland syndrome in Indonesia (Gupta & Kalra, 2021; Fiterman Rodrigues et al., 2020; Sardescu et al., 2017). Generally, Poland syndrome is characterized by hypoplasia or aplasia of the pectoral musculus and mammae hypoplasia and is associated with anomalies in the ipsilateral region of the body. The severity of limb anomalies can vary, ranging from syndactyly to phocomelia. Most cases are sporadic, but familial cases with intrafamilial variability have also been reported (Sardescu et al., 2017; Hashim et al., 2021).

The etiology of this syndrome is still unclear, but several hypotheses have been proposed (Buonsenso et al., 2022; Cui et al., 2024; Kamran et al., 2022). The generally applicable theory describes the disturbances at the end of the sixth to seventh weeks of pregnancy caused by an impaired embryonic blood supply resulting in hypoplasia of the ipsilateral subclavian artery. Some of the things that can cause this to happen include prenatal exposure to the potential of teratogens, such as cocaine, misoprostol, and smoking (Gupta & Kalra, 2021; Gui et al., 2018). The vascular disorder theory, the "subclavian artery supply disruption sequence" is the most acceptable pathogenesis mechanism (Hashim et al., 2021).

Poland syndrome also has a risk for the occurrence of intraoperative malignant hyperthermia, which is caused by the involvement of impaired musculoskeletal system development (Gupta & Kalra, 2021; Jun et al., 2023). Malignant hyperthermia is a critical

condition characterized by a rapid hypermetabolism of the skeletal muscle and an increased body temperature in response to certain volatile anesthetic agents and muscle relaxants, such as succinylcholine (Yang et al., 2020; Hopkins, 2011; Ellinas & Albrecht, 2020). For this reason, some literature suggests avoiding the use of anesthetic agents, such as inhaled anesthetics, especially halothane or depolarizing neuromuscular blockade agents, in patients with Poland syndrome (Fiterman Rodrigues et al., 2020; Díaz-Crespo et al., 2017). Since patients with Poland syndrome have an increased risk of developing malignant hypertrophy, special considerations are required, especially in terms of perioperative anesthesia management and the selection of appropriate general anesthetic agents. Some intravenous anesthetic agents have neuroapoptosis responses, such as ketamine, propofol, and thiopental. However, other anesthetic agents, such as midazolam and N2O, do not cause neuroapoptosis (Sabourdin et al., 2015). The continuous use of propofol in pediatrics is also associated with the occurrence of propofol infusion syndrome (Sabourdin et al., 2015). In a case report by Diaz-Crespo, the use of intravenous propofol in patients with Poland syndrome is associated with the onset of muscular spasms, which lead to the use of another sedation agent, that is, intravenous midazolam (Díaz-Crespo et al., 2017). This case report will explain the management of general anesthesia for a syndactyly release surgery in children with Poland syndrome.

This case report aims to describe the anesthetic management of a pediatric patient with Poland syndrome undergoing syndactyly release surgery, with a focus on preventing malignant hyperthermia through the use of total intravenous anesthesia. The report also aims to evaluate the clinical outcomes and safety of using midazolam-based intravenous anesthesia in this rare patient population. This case report offers both theoretical and practical benefits. Theoretically, it contributes to the limited literature on anesthetic management in pediatric Poland syndrome, supporting total intravenous anesthesia with midazolam as a safe alternative to volatile anesthetics in malignant hyperthermia-risk patients. Practically, it provides guidance for anesthesiologists and surgeons on preoperative assessment, malignant hyperthermia preparation, and anesthetic agent selection. The detailed technique, monitoring, and outcomes serve as a clinical reference for future cases and highlight the need for increased awareness of Poland syndrome among Indonesian healthcare professionals.

METHOD

Case Report

Clinical History

A 1-year- and 11-month-old boy came to the hospital with a diagnosis of Poland syndrome with a defect in the pectoralis major extra, syndactyly manus extra. He was scheduled to undergo syndactyly release surgery of interphalangeal manus dextra with a split-thickness skin graft. The patient's parents said that the fingers of the patient's right hand were fused together from birth. In addition, they also said that the shape of the patient's right chest had been slightly inward since birth. The patient was the first child and born through cesarean section on indications of a premature rupture of the membranes. A history of blue body and shortness of breath was not found. In familial anamnesis, no family experiences a similar condition. After explaining the action, the patient's family agrees and fills out the informed consent form.

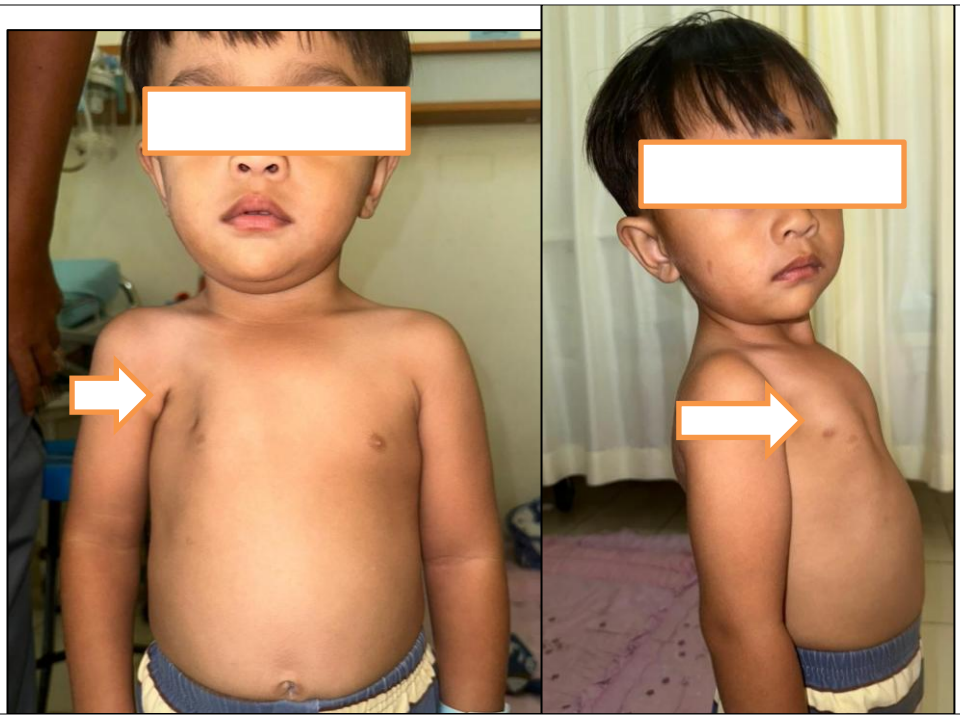

Figure 1. Patient clinical photos. A picture of hypoplasia in the pectoralis major dextra appears. The photo was taken with the consent of the patient's family.
Source: Patient medical record, taken with family consent

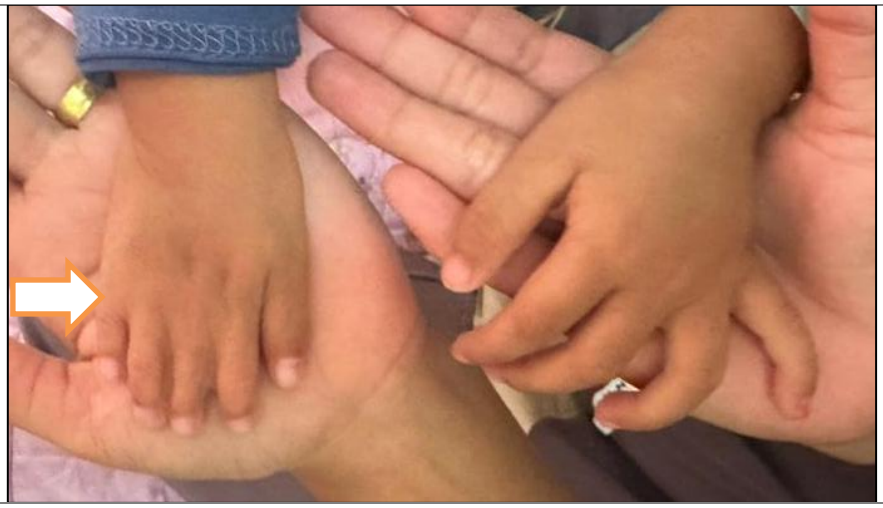

Figure 2. Patient clinical photos. There is a syndactily in the picture of the manus dextra. The photo was taken with the consent of the patient's family.
Source: Patient medical record, taken with family consent

Physical Examination

On physical examination, the patient was compos mentis, with a body weight of 11.1 kg and a height of 86 cm. The vital signs were found to be a pulse rate of 120 times/min, breathing rate of 30 times/min, saturation of 98% with room air, and body temperature of 36.8°C. The examination of the head found normocephal, conjunctiva, not anemical, sclera, not ecteric, and no mass in the neck. The examination of the thorax found abnormalities of the right chest wall, no pectoralis major (Figure 1). Paradoxical breathing movements are observed during deep inspiration. Vesicular lung sounds were observed in both pulmonary

fields, as well as regular heart sounds. No additional heart sounds were found. Abdominal examination found normal peristalsis. On the extremities, the fingers of the right hand are found united (syndactyly manus dextra) (Figure 2).

Table 1. Laboratory Examination Results

Parameters (units)	Results	Reference value
Hemoglobin (g%)	12.5	11.3–14.1
Leukocyte (/µL)	9400	6000–17,500
Thrombocyte (/µL)	357000	150,000–450,000
Prothrombin time (s)	14.3	11.6–14.5
International normal ratio (s)	1.00	<1.2
Activated partial thromboplastin time (s)	32.5	28.6–42.2
Blood sugar (mg/dL)	92	<200
Urea (mg/dL)	17	10.7–38.5
Creatinine (mg/dL)	0.39	0.39–0.55
Sodium (mmol/L)	141	135–145
Potassium (mmol/L)	4.2	3.5–5.5
Chloride (mmol/L)	109	97–107
Aspartate Transaminase	31	10–40
Alanine Transaminase	10	10–40
Albumin	4.7	3.8–5.4

Source: Patient medical record

Laboratory and Radiology Examination

The results of the laboratory examinations are shown in Table 1. During the X-ray examination, the lung and heart impressions were within normal limits (Figure 3).

Anesthesia Management

Before surgery, the patient was given pre-medication with intravenous midazolam 0.05 mg/kg. Before surgery, the pre-ductal and post-ductal oxygen saturation was 98%, with a heart rate of 120 times/min. Precautions to avoid malignant hyperthermia were prepared. Cooling liquid and cooling blankets were also prepared. Monitoring in the operating room used non-invasive blood pressure measuring devices, pulse rate, electrocardiography, and pulse oxygen saturation. The anesthesia machine was prepared without the use of an inhalation agent. A new circuit was used, and an intravenous 24G cannula was installed on the back of the patient's left hand.

The anesthesia team decided to proceed with total intravenous anesthesia using midazolam (0.1 mg/kgbw), fentanyl (2 mcg/kg), and atracurium (0.5 mg/kgbw) as anesthetic induction. Intubation was performed using GlideScope™ with blade size number 2 and continued with placing a 5.0 mm endotracheal tube. Anesthesia was maintained with continuous midazolam (0.3 mg/kg/hour) and fentanyl 1 mcg/kg intravenously administered intermittently. Measurement and monitoring of the patient's body temperature and end-tidal CO2 (EtCO2) were continuously conducted. After intubation, ventilation settings were performed with controlled ventilation modes (P insp 12 cmH2O, RR 26 times/min, I:E 1:2, PEEP 4 cmH2O) given, and EtCO2 was maintained at approximately 37–38 mmHg. The nasopharyngeal temperature was monitored at 36.0°C–36.5°C. A warming blanket and a blood warmer were fixed to ensure a suitable body temperature for the patient. Paracetamol,

tramadol, dexamethasone, and intraoperative ondansentron were given. The total anesthesia time was 120 min, with stable hemodynamics (Figure 4).

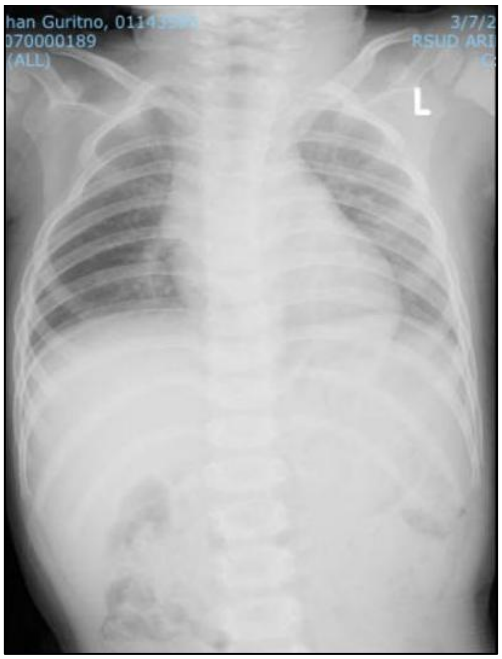

Figure 3. Patient's X-ray photo
Source: Patient medical record

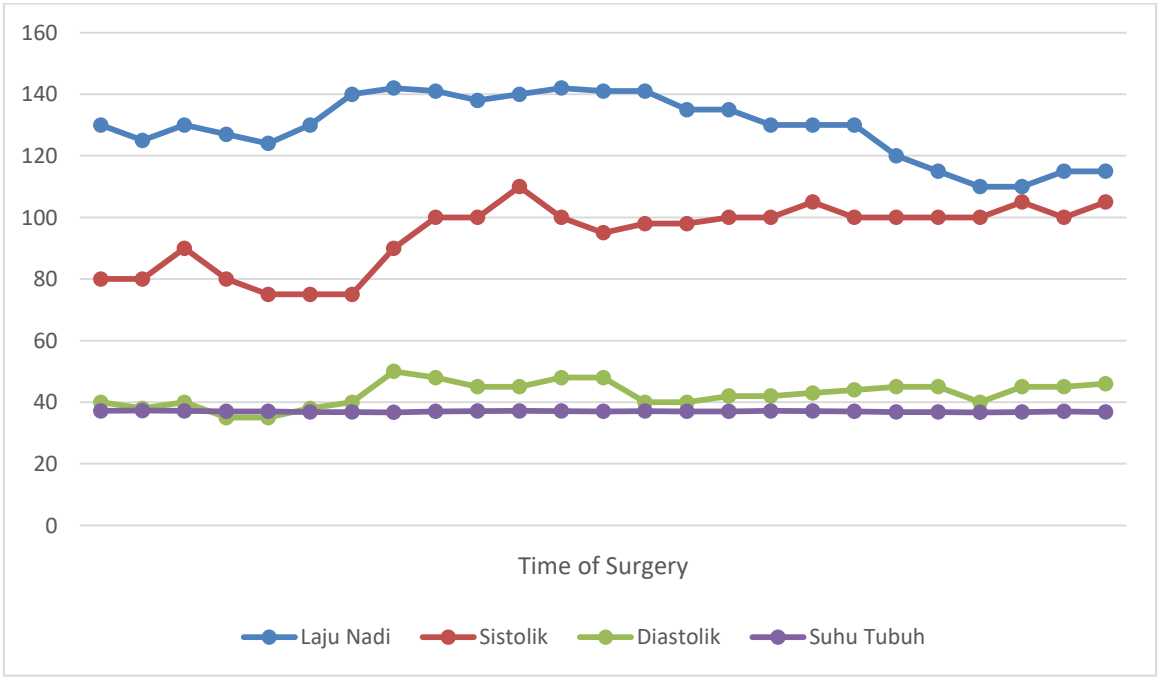

Figure 4. Intraoperative hemodynamic monitoring
Source: Patient medical record

The patient was extubated in the operating room, and then transferred to the recovery room for monitoring. No complications occurred during this procedure. After 60 min of monitoring in the recovery room, the patient was transferred to the treatment room.

Painkillers were given, that is, paracetamol 150 mg every 6 h on the first day. The patient was discharged after 2 days of treatment.

Table 2. Typical anomalies of Poland syndrome15

Musculoskeletal	Osteoarticular	Cardio Pulmonal	Others
Hypoplasia or absence of pectoralis major, absence of sternocostal part of the pectoralis muscle, rib hypoplasia, agenesis of the mammae glandular	Ipsilateral syndactyly, agenesis of the costal arch (pectus carinatum), scoliosis, hypoplasia of the bones of the arm and hand	Pulmonary bulbs (pneumothorax), postoperative atelectasis, dextrocardia, interventricular septal defects	Renal aplasia, hypospadia, ectopic testicles, leukemia, inguinal hernia, limited vascular access, Möebius syndrome (airway difficulties)

Source: Adapted from Díaz-Crespo et al. (2017)

RESULT AND DISCUSSION

Poland syndrome was first described by Alfred Poland about 180 years ago. The description is based on an autopsy on George Elt, a 27-year-old former inmate. Alfred Poland described 'pectoral muscle deficiency' in his autopsy report, which refers to underdeveloped anterior serratus muscles, brachysyndactyly in the ipsilateral hand, and abnormally small thoracic blood vessels. In addition, there is also hypoplasia of the pectoralis major muscular in the sternum and costal part and the absence of the pectoralis minor muscular disease.4

Until now, the pathophysiology and etiology of Poland syndrome are still unclear. There are many theories about its pathophysiology, but the most widely accepted theory is that there is a vascular defect in the subclavian artery that occurs on the 45th day of embryonic life, where the vascular defect disrupts the development of body parts in the scapulo-humeral region. This results in an insufficient flow in the body, chest, and distal extremities, ultimately giving rise to Poland syndrome's main musculoskeletal characteristic (Baldelli et al., 2020).

There is no definitive treatment of Poland syndrome yet. Surgical procedures are generally necessary to correct primary defects and other related medical problems. These procedures are usually not too different from those of other similar malformations in the general population (Baldelli et al., 2020).

Patients with Poland syndrome with thoracic abnormalities (especially children) may experience airway difficulties at the time of intubation under general anesthesia. Therefore, preoperative radiological imaging should be used to prepare for adequate anesthesia treatment to determine a tracheal deviation or stenosis and evaluate respiration-related chest movements (Jun et al., 2023). In this case, the patient experienced several chest wall abnormalities, but the lungs on the right side were considered normal, and there were no abnormalities in the lung function. Therefore, problems related to the lung function during general anesthesia are considered minimal, and the administration of anesthesia is more emphasized in the estimation of the development of malignant hyperthermia. Malignant hyperthermia is an additional risk given congenital musculoskeletal changes, although few publications support the fact of a link between Poland syndrome and malignant hyperthermia (Díaz-Crespo et al., 2017).

Table 3. Advantages and disadvantages of total intravenous anesthesia in pediatrics16

Advantages
● Reduces the risk of postoperative nausea and vomiting
● Reduces room pollution
● Reduces the incidence of delirium when recovering consciously
● Reduces the likelihood of laryngospasm and bronchospasm
● Intravenous total anesthesia is highly recommended in neuromuscular diseases, myopathy, and muscular dystrophy
● Less interference with evoked potential monitoring
Disadvantages
● Need for venous access and pain at the time of injection
● Risk of bacterial contamination
● Current inability to monitor concentrations in the blood
● Need for special tools (e.g., infusion pumps)
● Need for special training for intravenous total anesthesia techniques
● Potential and risks of intraoperative awareness
● Monitoring the depth of anesthesia in young children is less likely to be reliable

Source: Adapted from Anderson & Bagshaw (2019)

Patients who are at risk of developing malignant hyperthermia should avoid exposure to inhaled anesthesia or suxamethonium (Hopkins, 2011). All inhaled anesthetics, such as halothane, ether, desflurane, sevoflurane, and isoflurane, except for nitrous oxide, have been reported as triggering malignant hyperthermia. The neuromuscular depolarizing inhibitor agent succinylcholine is associated with the side effects assessed to induce malignant hyperthermia. All anesthetic agents other than volatile anesthetic agents and non-depolarizing neuromuscular inhibitors are considered safe from malignant hyperthermia. In addition to the problem of volatile agents, Poland syndrome itself can also increase the risk of hyperthermia maligna (Jun et al., 2023). In conditions where general anesthesia is required, strategies to avoid malignant hyperthermia-inducing substances, such as inhaled anesthetic agents and depolarizing neuromuscular inhibitors, are essential (Hopkins, 2011). Monitoring body temperature and end-tidal CO2 is also necessary in monitoring and checking the occurrence of malignant hyperthermia (Jun et al., 2023).

Potential inhaled anesthetic agents can be substituted in patients at risk of malignant hyperthermia, who require general anesthesia using the technique of total intravenous anesthesia (Hopkins, 2011). The common anesthetic agents used in intravenous total anesthesia in pediatric patients are propofol, midazolam, ketamine, dexmedetomidine, and opioids (Cowie et al., 2019). Intravenous midazolam is an excellent sedation and anxiolytic agent with a more stable hemodynamic effect when compared to propofol (Cowie et al., 2019). The initial dose used was 0.05–0.1 mg/kgbw intravenously, with a maintenance dose of 0.1–0.3 mg/kgbw intravenously (Course & Mogane, 2022; Romanini et al., 2018). The intravenous use of midazolam is associated with decreased serum cytokine levels and may modulate pre-existing proinflammatory status in pediatric patients and prevent suppression of the immune system resulting from anesthesia and surgical procedures (Lotfy et al., 2021). In a report by Diaz-Crespo, the use of intravenous propofol in patients with Poland syndrome was associated with the onset of muscular spasms, which led to the use of another sedation agent, called intravenous midazolam (Díaz-Crespo et al., 2017). The use of propofol in pediatrics is also associated with the occurrence of propofol infusion syndrome and

neuroapoptosis response (Sabourdin et al., 2015). Due to these considerations, in this case, the patient decided to use intravenous total anesthesia using the anesthetic agent midazolam.

Patients with Poland syndrome require a detailed physical examination as chest wall abnormalities and accompanying malformations can occur. In addition, it is important to anticipate the problem and recognize the associated abnormalities beforehand. When administering anesthesia to a patient with Poland syndrome, the anesthesiologist must be fully aware of the risk factors for malignant hyperthermia and the accompanying lung problems. During the intraoperative and postoperative periods, no complications occurred in these patients. After 2 days of treatment, the patient was discharged without complications.

CONCLUSION

The primary principle of anesthesia management in patients with Poland syndrome is the prevention of intraoperative malignant hyperthermia, a potentially life-threatening complication that can lead to severe morbidity or death; therefore, careful preventive strategies must be implemented. In this case, the use of total intravenous anesthesia with midazolam for the surgical release of manus dextra interphalangeal syndactyly in a child with Poland syndrome proved to be a safe and clinically beneficial option. Although anesthesia management in patients with Poland syndrome remains challenging, thorough preoperative assessment, meticulous preparation, and well-planned intraoperative management are essential to ensure safe anesthetic care. Anesthesiologists must remain vigilant regarding possible complications associated with this rare condition. Future research with larger case series or multicenter studies is recommended to further evaluate the safety and effectiveness of total intravenous anesthesia techniques in pediatric patients with Poland syndrome.

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