DUCTAL STENTING IN PULMONARY ATRESIA NEONATES WITH MULTIPLE CONGENITAL ANOMALIES AND SEPTIC CONDITION

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ABSTRACT

Neonates with pulmonary atresia usually appear normal at birth with pulmonary circulation maintained by the presence of a patent ductus arteriosus (PDA). Rapid deterioration will suddenly occcur if the duct close. Surgical shunt is still be used as a standard protocol in many centers as a palliative procedure. We report a 2 days-old, low birth weight, and mild cyanotic neonate with pulmonary atresia and PDA accompanied by atresia ani, bladder and cloaca extropy, ambiguous genitalia and sepsis. We decided to perform PDA stenting because our patient have a high surgical shunt risk. This procedure was very important to keep the duct remains open until patient ready for total surgical correction. **[MEDICINA 2015;46:42-45].**

Keywords: *ductal stenting, ductal dependent pulmonary blood flow, pulmonary atresia.*

PEMASANGAN *STENT* DUKTUS ARTERIOSUS PADA NEONATUS DENGAN ATRESIA PULMONAL, KELAINAN KONGENITAL MULTIPEL, DAN SEPSIS

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ABSTRAK

Neonatus dengan atresia pulmonal biasanya tampak normal saat lahir dengan adanya *patent ductus arteriosus* (PDA) yang memelihara aliran darah paru. Kondisi neonatus akan segera memburuk jika duktus menutup. Pembuatan *shunt* dengan pembedahan merupakan protokol standar yang masih dikerjakan di banyak pusat kesehatan. Kami melaporkan neonatus berusia 2 hari dengan berat badan lahir rendah dan sianosis ringan dengan diagnosis atresia pulmonal, PDA, atresia ani, ekstropi buli-buli dan kloaka, jenis kelamin ambigu, dan sepsis. Kami memutuskan untuk melakukan pemasangan *stent* pada PDA karena pasien kami memiliki risiko yang tinggi untuk pembedahan (pembuatan *shunt*). Tindakan ini sangat penting untuk menjaga duktus tetap terbuka sampai pasien siap untuk dilakukan operasi koreksi. **[MEDICINA 2015;46:42-45].**

Kata kunci: *stent duktus arteriosus, aliran darah paru tergantung duktus, atresia pulmonal.*

INTRODUCTION

In pulmonary atresia, there is no communication between the right ventricle and the pulmonary artery so that a patent ductus arteriosus (PDA) or no collateral arteries become a major source of blood flow to the lungs. Venous blood flow entering the right atrium to the left atrium through an atrial septal defect or a patent foramen ovale (right to left shunt). Right atrium will be dilated and right ventricle maybe hypoplastic or normal. If the pulmonary blood flow is only maintained by the collateral arteries or if the PDA closes early after birth, then there would be reduced pulmonary blood flow and neonates will experience cyanosis.[1]

Neonates with ductal dependent pulmonary blood flow would benefit from maintained ductal patency for several months until he is ready to undergo definitive surgery. Conventional palliative therapy to maintain pulmonary blood flow is to provide a prostaglandin E1 infusion until ready to palliative aortopulmonal shunt (Blalock-Taussig shunt or BT shunt).[2] Prostaglandin has side effects such as peripheral vasodilation, hypotension, hyperpyrexia, and can cause apnea.[3] Modified BT-shunt during postnatal period has a high mortality risk dan complication in example chylothorax, vagus and phrenicus nerve paralysis, early or

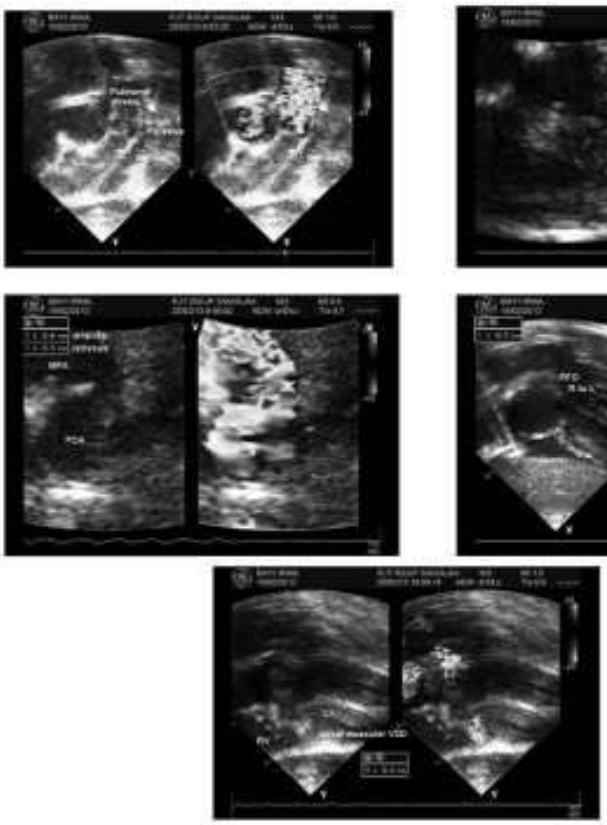

Figure 1. Echocardiography results.

late shunt stenosis, distortion and imbalance pulmonary artery growth, adhesion due to surgery.[2-4] We described a case of pulmonary atresia with successful PDA stenting as an alternative to maintain pulmonary blood flow.

CASE ILLUSTRATION

A 2 days-old mild cyanotic low birth weight baby (SpO2 89-90%, 2050 grams) reffered to our hospital due to multiple congenital anomalies accompanied by suspicion of congenital heart defects of PDA. The baby was a second child from a 29-year-old mother (G_2P_{1001}), term infants (37-38 weeks), through a process of spontaneous labor in midwives and said to burst into tears. Physical examination did not show signs of severe cyanosis of the mucosa and fingers. There was a continuous murmur heard at the upper left parasternal line (second intercostal) with a single second heart sound are single not harden. His reddish intestine were spilled out without membrane, the baby's gender can not be determined, atresia ani with faeces came out from the external genitalia. Laboratory findings showed an increase markers of infection and further deterioration occurs.

Echocardiography showed a pulmonary atresia (with native pulmonary valve), hypoplastic right ventricle, moderate tortuous PDA (isthmus 3 mm and ampulla 4 mm, left-to-right shunt), a-3 mm persistent foramen ovale with right to the left shunt, a-2 mm apical muscular ventricular septal defect with left to right shunt, minimal pericardial effusion, with good left ventricular function (ejection function 82%) (**Figure 1**). Therapy was subsequently given to these patients was 10 mcg misoprostol every 6 hours, and advised not to use oxygen

Ductal stenting was performed at the age of 17 days. The first 3.0/9 mm BMS (Bare metal stent) and the second 2.75/9 mm BMS were delivered via a right Judkins catheter 4/5F and fully covered the PDA (**Figure 2**).

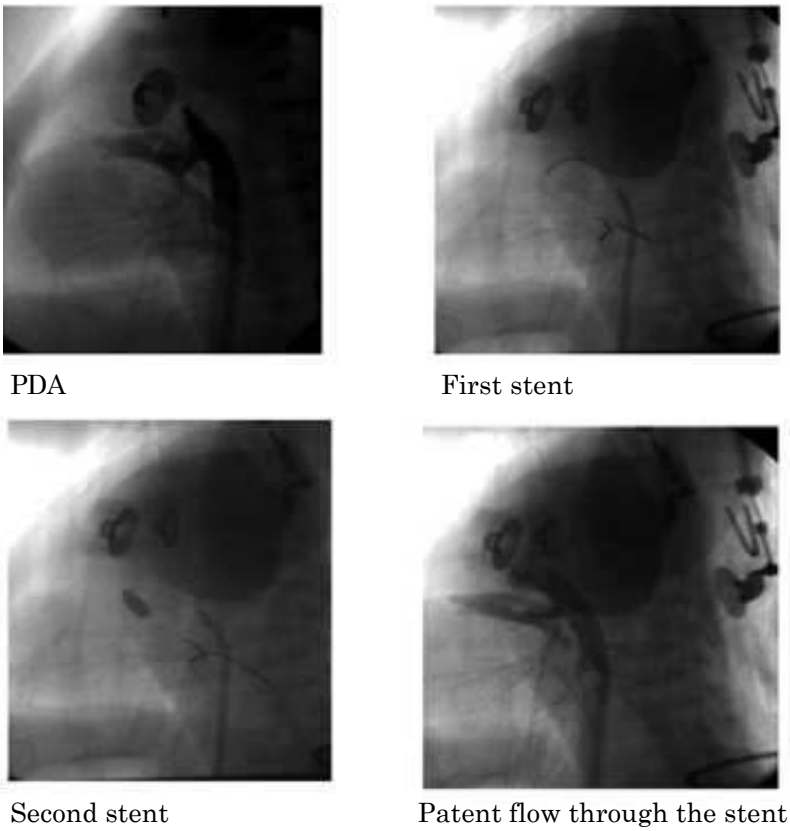

Figure 2. Ductal stenting intervention procedure.

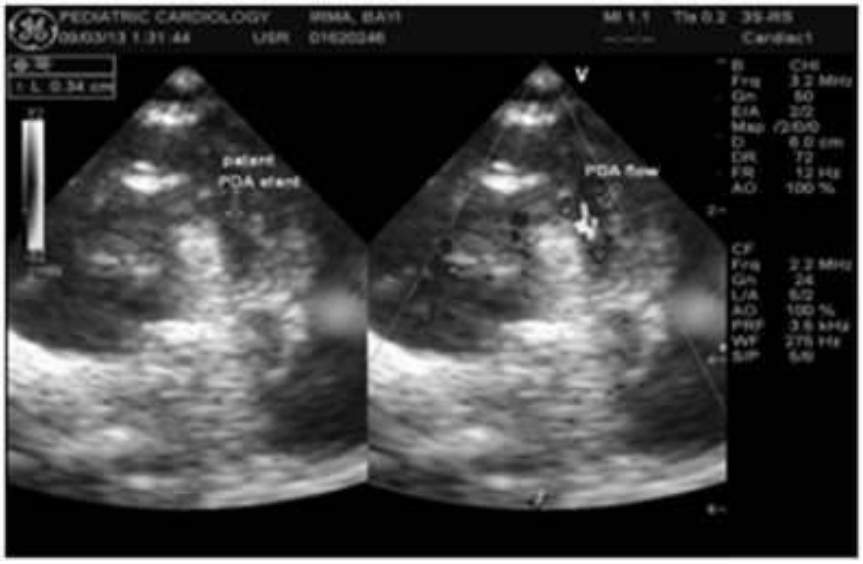

Figure 3. Echocardiography evaluation showed patent stent.

Oxygen saturation was dropped to 30% during inflation and went back to 89% after deflation. Evaluation after stenting showed patent flow through the PDA. Heparin 10 unit/kgBW/hour was given after the procedure.

The baby experienced desaturation (peripheral saturation of 60%) six hours after stenting. Echocardiography evaluation showed that the PDA stent was patent (**Figure 3**). Two days after stenting, the baby died due to sepsis.

DISCUSSION

Ductal stenting in cyanotic congenital heart disease neonates with duct dependent pulmonary blood flow could be an alternative to palliative surgical intervention especially for high risk patients such as premature babies or low birth weight babies. Originally they used early generation rigid stent with a stiff and thick wire which was easily cause complications including worsening cyanosis, ductal spasm, bleeding, rupture of the blood vessels, tissue prolapse, acute thrombosis, and peripheral arterial disorders.[2]

Patent ductus arteriosus has two edges, the narrowest part called isthmus and edges from the aorta which called ampula. Patent ductus arteriosus has a unique structure, the tunica media is mainly composed of muscle tissue and the intima is thicker than other blood vessels. This structure makes PDA easily spasm.[4,5] Anatomy of the duct affects the success of stenting. Horizontal and tubular duct has a higher success rate for stenting. Ductal stenting primary indication is when neonates are at high risk for surgery, unavailability of adequate surgical facilities, ideal ductal morphology (tubular/ straight duct), the need to maintain ductal patency for more than 2 weeks. Tortuous duct and univentricular heart are avoided to stent and only done stenting if there are limitations to perform palliative surgery.[6] In our case, we sucessfully deploy two stents in a tortuous duct in which our patient is not suitable to undergo palliative surgery due to low birth weight.

Several studies recommend to discontinue prostaglandin infussion 4-6 hours before stenting so that the duct can hold the stent.[5,7-9] Others recommend to stopped prostaglandin at the time of procedure when the duct has been passed by the guiding wire in severe cyanosis neonates with the duct started constricting and very depend on the duct or turtuous duct.[2,10] We used syntetic prostaglandin orally because prostaglandin infussion was not available in our hospital and we stop prostaglandin just before the procedure.

It is recommended to use a long, flexibel, low profile ballon stent.[2,11] Stent lenght selected based on ductal anatomy and size. Lenght measurement are made when the wire already inside the PDA.[5,6] The stent should cover the entire duct from the aortic edge to the pulmonic edge, because the distal side of duct is easily constrict. It is recommended to deploy only one stent whenever possible.[2] We deploy two stent in our case because there was a limitation to measure the ductal lenght due to its tourtous ductal morphology and succesfully succesfully cover the entire duct.

Stent diameter 3-4 mm and post stenting ductal diameter 4-5 mm could reduce the heart work and regulate pulmonary blood flow.[2,4] Stenting of the duct provides pulmonary artery development thus it giving more time to prepare definitive surgery. Stenting success was marked by an increase of oxygen saturation, improvement of PDA diameter and cyanosis, and decrease in diastolic blood pressure.[9] Our patient's oxygen saturation was significantly decrease during ballon inflation and then back to 89%. This was in accordance with previous reports.

Ductal stenting could reduce hospital lenght of stay, the babies can safely going home in 48 hours after procedure. Evaluation usually done 6-12 weeks after that until definitive surgery performed.[5] Surgery after stenting is safe and has a low risk.[3] Our patient was still being treated in intensive ward for other reason and unfortunately died due to worsening of septic condition.

SUMMARY

Our case demonstrate a low birth weight neonate who was suffered from pulmonary atresia with PDA, multiple congenital

anomalies, and also in the septic conddition. The suspicion of pulmonary atresia and PDA were made by finding of mild cyanotic baby with single second heart sound, and continuous murmur. The diagnosis was confirmed by echocardiography examination. We have done PDA stenting with two stent as an alternative to surgical shunt for our high risk patient (low birth weight baby) to maintain pulmonary blood flow. Although we successfully done PDA stenting, the condition of the patient did not show satisfying progress due to septic condition.

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