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經由導管技術對於肺動脈不全病人肺動脈成長及開刀治療結果之影響

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中文摘要：

背景：

肺動脈閉鎖常合併肺動脈發育不全, 或甚至嚴重畸型造成日後開刀矯正的困難。傳統的開刀手術需多次的分流手術及側枝循環的結紮，近來心導管術的進步，可利用心導管做肺動脈復健。(transcatheter pulmonary artery rehabilitation)本研究評估該技術對於肺動脈成長的效果，及臨床追蹤或開刀之結果。

方法：

1. 對於分流管(Goretex shunt)狹窄者，以氣球導管擴張，若合併肺動脈狹窄，則再做肺動脈擴張術。
2. 接受 bidirectional Glenn shunt 的病人則栓塞奇靜脈。
3. 側枝循環存在的病人，則以螺旋線圈栓塞術栓塞不必要的側枝循環。
4. 比較術前，術後之臨床狀況包括 a. 心臟衰竭之有無。b. Mcgoon Index。c. 開刀結果。

研究對象：

從 1996 年至 2003 年間，共有 119 位病人進入本研究。病人分兩組，一組為接受分流管或及肺動脈張術(95 人)，另一組為接受側枝循環或奇靜脈栓塞術(24

人)。

結果：

第一組病人分流管狹窄病人 95 人，合併肺動脈狹窄者有 32 人，擴張分流管 89 人(94%)，失敗 6 例，肺動脈狹窄擴張 31 人，失敗 1 例。89 例擴張分流管病人，術後血中氧氣濃度增加 3%以上者有 85 例，血中氧氣飽和度由平均 73 ± 7%，增加為 78 ± 6% (P < 0.01)肺動脈擴張術增加最窄部份增加 50%以上的 diameter 病人有 22 人(22/31, 71%)，經平均 24.1 ± 11.8 月追蹤後，平均 McGoon ratio 由 1.3 ± 0.3 增加至 1.4 ± 0.4。(P < 0.01)

第二組病人 24 例接受側枝循環栓塞, 其中 21 例有動脈分枝形成(arborization)的異常，栓塞 21 例在 MAPCA，3 例在奇靜脈(a2ygos vein)栓塞在 24 例均獲成功氧氣飽和度由平均 86 ± 4%降至 84 ± 5%，在一至三月後追蹤氧氣濃度為 85 ± 5%，但病人心臟衰竭症狀顯著改善有 23 例(3 例術後可以成功拔管)，術後六個月以電腦斷層評估，平均 McGoon ratio 由 1.2 ± 0.3 增至 1.4 ± 0.4，矯正手術有 10 人成功，手術後不幸死亡 2 例，有 12 人當在追蹤。

結論：

經由心導管的肺動脈復健技術, 對於肺動脈閉鎖病人肺動脈的成長有顯著的效果，因此成為治療之首選。

Abstract

Background:

Pulmonary atresia with ventricular septal defect (PA-VSD) is frequently associated with hypoplasia of central pulmonary artery which prevents successful repair. Unifocalization and a shunt is a classic therapeutic approach. Transcatheter rehabilitation has recently applied in the treatment of PA-VSD.

Method:

Balloon angioplasty was performed in stenotic Goretex shunt and pulmonary arteries. Embolization of MAPCA or a2ygos vein was performed following unifocalization & shunt to redirect the flow to PA.

Patients:

Between 1996 and 2003, 119 patients were enrolled in this study. Two groups were identified. Group I consisted of 95 patients with stenosis in shunt or pulmonary arteries. Group II consisted of 24 patients with MAPCA or a2ygos veins.

Results:

In group I, balloon angioplasty for stenotic shunt was successful in 89(94%), but failed in 6. Angioplasty for a concomitant pulmonary artery stenosis was performed in 31 and failed in 1. The O_2 saturation increased from a mean of 73 ± 7% to 78 ± 6%. ($P<0.01$) pulmonary artery angioplasty was effective in 22 (71%). The McGoon ratio increased from 1.3 ± 0.3 to 1.4 ± 0.4. In group II, 21 patients had MAPCA and 3 had a26gos vein. Embolization of MAPCA or a2ygo vein was successful in ale. O_2 saturation declined from 86 ± 4% to 84 ± 5%. But increased to 85 ± 5% at 1~3

months follow-up. Heart failure symptom improved in 23. The McGoon ratio at 6-months increased from a mean of 1.2 ± 0.3 to 1.4 ± 0.4. In group II, a definitive repair was performed in 10 with 2 mortality.

Conclusion:

Transcatheter rehabilitation is quite effective in facilitating growth of hypoplastic central pulmonary artery and should be considered as a conjunctive treatment in pulmonary atresia complex.

Introduction:

Pulmonary atresia and ventricular septal defect is frequently associated with hypoplasia, absence or arborization anomaly of central pulmonary artery which prevents successful repair. The classical management of patients with complex pulmonary atresia required repeated surgery including unifocalization and ligation of MAPCAS to facilitate growth of pulmonary arteries before repair of intracardiac defects. Recently transcatheter technique was applied for pulmonary artery rehabilitation, which resulted in better growth of pulmonary arteries and therefore, operation results were improved. We assess the efficacies of this technique to the growth of pulmonary arteries in this group of patients. The outcome and results of surgeries were also evaluated.

Methods & Patients:

Hemodynamic studies were routinely performed after insertion of an appropriate size sheath in femord artery. Angiography was performed in the subclavian artery or in the shunt to detect stenosis in shunt or pulmonary artery. Balloon angioplasty was performed in patients who had stenosis in Goretex shunt and pulmonary artery. The balloon used was generally 20% large than diameter of the Goretex shunt. O_2 saturation in aorta was analyzed prior & 15 minutes following the procedure. The method of dilation has been described in a previous report. The O_2 saturation in the aorta was measured before & 15 minutes after the procedure. Patients with massive MAPCA received angiography to delineate the distribution & morphology of MAPCAS. Unifocalization or a shunt was performed to facilitate the growth of pulmonary artery as a primary procedure. Following a shunt or unifocalization, transcatheter closure of MAPCA was performed in an interval of 2 weeks to 3 months. For those with MAPCAS, coil embolization was performed to eliminate dual supply.

The coil dimension selected was generally 2 times of minimal diameter of MAPCA. The coils were delivered via an end-hole catheter via a retrograde aortic root. AS many as coils were deloyed to achieve complete embolization artery was detected in these 32 patients. We evaluate the effectiveness of transcatheter rehabilitation in terms of symptoms and pulmonary artery size.

Results:

Between a 7-year period, 119 patients with associated pulmonary atresia or severe infundibular stenosis were enrolled in this study. Two groups were identified. Group I consisted of 93 patients with stenosis in modified shunts and/or pulmonary artery. Of the 93, 32 patients had associated pulmonary artery stenosis or forty-one stenoses in pulmonary arteries. Angioplasty for stentic shunt was successful in 89 patients, but failed in 4. Of the 89 patients, the mean O2 saturation increased from 73 ± 7% to 78 ± 6%. A simutaneas dilation of stenotic pulmonary artery was successful in 22(71%) patients. After a mean follow-up period of 24.1 ± 11.8 months, the mean McGoon ratio increased from 1.3 ± 0.3 to 1.4 ± 0.4. ($P<0.01$) The Group II consisted of 24 patients. There were 21 patients with MAPCAS and arborization anomaly in pulmonary arteries. The O_2 saturation decreased from a mean of 86 ± 4% to 84 ± 5%. ($P<0.01$) Symptomatic improvement was documented in 23. All 24 patients had symptoms of heart failure. Three requiring respiratory assist could be extubated and weaning of respirator was successful in these 3 patients within 7 days. The follow-up CT imaging showed the McGoon ratio increased from 1.2 ± 0.3 to 1.4 ± 0.4.($P<0.01$) A Rastelli type operation was performed in 12 patients with 2 mortalities: 1 early mortality and 1 late mortality. Of the latter case, the mortaligy was incriminated to airway obstruction in an episode of Bronchiolitis. The remaing 12 patients are awaiting a definitive surgery. One of the 12 patients had extremely

diminutive pulmonary artery size that a total repair is considered as not feasible.

Three undergoing bidirectional Glenn shunt had a2ygo vein which directed blood flow to inferior vena cava. Embolization of MAPCA and a2ygos vein with coils performed in 21 and 3 patients, respectively (Figure2).

Discussion:

Pulmonary atresia with ventricular septal defect of tetralogy of Fallot was difficult to be repaired. Management strategy included shunt and unifocalization in early infancy, ligation of MAPCA and Rastelli type operation if pulmonary artery size was inadequate. However, hypoplasia or agenesis of central pulmonary artery is frequently present in those patients rendering difficulties in definitive repair. Meanwhile, the technical difficulties & the need for insertion prothetic conduit to small peripheral artery were frequently encountered in unifocalization.

In a previous article, we have documented the effectiveness of balloon angioplasty for obstructed Goretex shunt and pulmonary artery stenosis. In this study, transcatheter balloon dilation was effective in the growth of pulmonary artery. Therefore, balloon dilation for obstructed Goretex shunt can be an alternative to a second shunt, if pulmonary artery size was suboptimal. Pulmonary artery stenosis either congenital or acquired is frequently seen is patients with pulmonary atresia. Balloon angioplasty could be effective in 50% cases.

MAPCA is frequently present in patients with arborization anomaly in pulmonary arteries, and blood flow was directed to branch pulmonary arteries by MAPCA. Central pulmonary arteries were generally hypoplastic. The MAPCA should be ligated if dual supply is present. In the presence of MAPCA, lung flow became excessive that plethorax is frequently present. Transcatheter embolization provided an alternative to ligation of MAPCA. A2ygos vein and collateral veins could developed

following bidirectional Glenn, where patients may develop profound cyanosis. Transcatheter embolization of the a2ygos vein and collateral veins could increase the pulmonary blood flow and hence increase systemic O_2 saturation.

A right ventricle-pulmonary artery (RV-PA) conduit was recommended in those with hypoplastic central pulmonary arteries. Following placement of RV-PA conduit, the growth of pulmonary artery could be achieved. The ventricular septal defect should be closed in a selected time.

A Rastelli type operation required adequate pulmonary artery index. Unifocalization or placement of RV-PA conduit may be useful in facilitating growth of pulmonary artery. Transcatheter balloon dilation for stenotic branch pulmonary artery could be a useful procedure. Coil embolization of MAPCA or a2ygos vein redirected the flow to central pulmonary artery which facilitated pulmonary artery growth. Therefore, we conclude that transcatheter rehabilitation is effective in facilitating the growth of pulmonary artery in complex pulmonary atresia.

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Figure Legend

Figure 1. An angiogram at left suhclavian artery at Goretex shunt following unifocalization. Following angioplasty the stenosis were relieved.

Figgure 2. An angiogram at SVC showing a2ygos vein diverting SVC flow to IVC. Following coil embolization , the a2ygos vein was occluded.

Figure 1

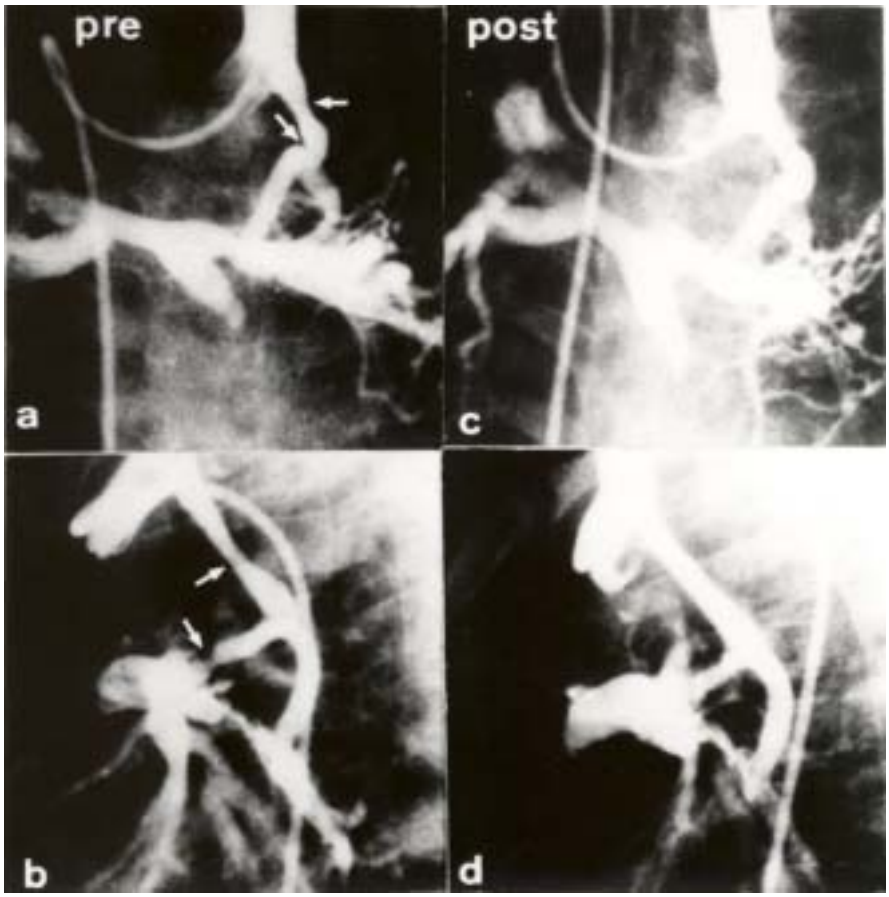

Figure 2