

成人左冠状动脉起源于肺动脉外科治疗



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【摘要】 目的 总结成人左冠状动脉起源于肺动脉的外科治疗结果及经验。方法 1991 年 11 月至 2017 年 11 月, 手术治疗 36 例年龄大于 18 岁左冠状动脉起源于肺动脉患者, 其中男 9 例、女 27 例, 年龄 (36.6±13.3) 岁, 体重 (60.0±9.4) kg。术前超声心动图显示左心室射血分数 (LVEF) 为 57%±6%, 左心室舒张期末内径 (LVEDD) 为 (52.3±6.3) mm; 二尖瓣反流 (MR) 重度 1 例, 中度 5 例。其中行冠状动脉再植 17 例, 肺动脉内隧道修补术 (Takeuchi 术) 16 例, 异常起源左冠状动脉结扎+冠状动脉旁路移植术 3 例, 同期行二尖瓣成形术 6 例。结果 体外循环时间为 (152.5±72.9) min, 主动脉阻断时间 (101.9±43.6) min, 无院内死亡, 呼吸机辅助时间 (17.3±16.3) h, ICU 滞留时间 (43.1±30.7) h, 术后 LVEF 为 59%±6%, 较术前无明显改善 ($P=0.10$), 术后 LVEDD 为 (46.9±5.9) mm 较术前显著缩小 ($P=0.02$), 6 例二尖瓣成形患者中, 1 例重度变为轻度, 5 例中度变为微量。所有患者顺利出院。随访 35 例, 平均 5.5 年, 最长 26 年, 随访期间无死亡, 2 例行肺动脉内隧道修补术患者发生肺动脉内隧道瘘, 分别成功行介入封堵术和外科修补术; 所有患者心功能分级 (NYHA) I 或 II 级; 最后一次随访 LVEF 为 69%±7%, 较术前显著改善, LVEDD 为 (48.7±5.9) mm; MR 中度 2 例, 轻度 10 例。结论 成人左冠状动脉起源于肺动脉外科治疗近期和远期效果满意, 肺动脉内隧道修补术远期可能出现内隧道瘘需要再次干预。

【关键词】 左冠状动脉起源于肺动脉; Takeuchi 术; 冠状动脉再植

Surgical treatment of adult patients with anomalous left coronary artery from the pulmonary artery

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【Abstract】 Objective To review the experience of the surgical treatment of adult patients with anomalous left coronary artery from the pulmonary artery (ALCAPA). **Methods** A retrospective, single institution review was conducted on thirty-six adult patients with ALCAPA surgical treatment from November 1991 to November 2017 in Fuwai Hospital. Of these patients, nine were males and twenty-seven were females. The mean age was 36.6±13.3 years. The mean weight was (60.0±9.4) kg. The preoperative echocardiography showed the mean left ventricular ejection fraction (LVEF) was 57%±6% and the mean left ventricular end-diastolic dimension (LVEDD) was 52.3±6.3 mm. Severe mitral regurgitation (MR) was seen in one patient, moderate in five patients. The operative procedures included coronary artery re-implantation in seventeen patients, Takeuchi operation in sixteen patients, ligation of left coronary artery plus coronary artery bypass graft in three patients. In addition, six patients underwent mitral valve repair. **Results** There was no in-hospital mortality. The mean cardiopulmonary bypass time was 152.5±72.9 min and cross clamp time was 101.9±43.6 min, respectively. The mean mechanical ventilation time and ICU time were 17.3±16.3 h and 43.1±30.7 h, respectively. The mean postoperative LVEF was 59%±6%, which did not significantly improve compared with preoperative LVEF. However, the mean postoperative LVEDD of 46.9±5.9 mm had significant reduction compared with the preoperative LVEDD. Of the six patients with mitral valve repair, one was mild and the other five were trivial. Thirty-five patients (97.2%) completed the follow up with a mean time of 5.5 years. All the patients survived with New York Heart Association class I or II. Two patients needed interventional occlusion or re-operation due to the fistula of internal tunnel within the

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pulmonary artery. At the latest echocardiography, the mean LVEF of $69\% \pm 7\%$ improved significantly compared with the preoperative LVEF. Mild MR was detected in ten patients, moderate in two patients during the follow-up period.

Conclusion The surgical treatment of adult patients with ALCAPA has satisfactory short-and long-term results. The patients underwent Takeuchi procedure may need re-operation due to fistula of internal tunnel within the pulmonary artery during the long follow-up.

【Key words】 Anomalous left coronary artery from pulmonary artery; takeuchi procedure; coronary re-implantation

左冠状动脉起源于肺动脉 (ALCAPA) 是一种罕见的先天性心脏病, 其发病率约为 1/300 000, 如不及时外科治疗, 1 年内自然死亡率可高达 90%^[1]。极少患儿由于左右冠状动脉形成良好的侧枝循环可以生存至成年, 由于发病率低和死亡率高的特点, 成人左冠状动脉起源于肺动脉在临床更为罕见, 仅少量病例报道^[2-3]。阜外医院成人外科中心 1991 ~ 2017 年共外科治疗成人左冠状动脉起源于肺动脉 36 例, 现将结果总结报道如下。

1 资料与方法

1.1 临床资料

1991 年 11 月至 2017 年 11 月, 阜外医院成人外科中心共收治 36 例年龄 > 18 岁左冠状动脉起源于肺动脉患者, 其中 2010 年以前完成 6 例, 2010 年以后完成 30 例。男 9 例、女 27 例, 年龄 (36.6 ± 13.3) 岁, 体质量 (60.0 ± 9.4) kg。首发症状为活动后胸闷或胸痛 29 例、心脏杂音 5 例、因行其他外科治疗术前检查发现 2 例。术前超声心动图、CT 或冠状动脉造影均提示左右冠状动脉之间侧枝循环丰富、右冠状动脉起始部增粗和迂曲。术前超声心动图显示左心室射血分数 (LVEF) $57\% \pm 6\%$, 左心室舒张期末内径 (LVEDD, 52.3 ± 6.3) mm, 二尖瓣反流 (MR) 重度 1 例, 中度 5 例。

1.2 手术和随访方法

其中行冠状动脉再植 17 例, 肺动脉内隧道修补术 (Takeuchi 术) 16 例, 异常起源左冠状动脉结扎+冠状动脉旁路移植术 3 例, 同期行二尖瓣成形术 6 例。成形技术包括 6 例患者植入二尖瓣成形环, 其中 3 例患者行乳头肌松解。所有手术均在全身麻醉低温体外循环下完成。术后通过门诊或电话随访。

1.3 统计学分析

采用 SPSS22.0 统计软件进行分析, 计量资料以均数±标准差 ($\bar{x} \pm s$) 表示, 采用 *t* 检验; 计数资料以率表示, 比较采用卡方检验; 以 $P < 0.05$ 为差异有统计学意义。

2 结果

所有患者均顺利出院, 无严重并发症发生 (再次气管插管、机械辅助、肾功能衰竭、严重感染、伤口愈合不良等)。体外循环时间 (152.5 ± 72.9) min, 主动脉阻断时间 (101.9 ± 43.6) min, 呼吸机辅助时间 (17.3 ± 16.3) h, 住 ICU 时间 (43.1 ± 30.7) h, 出院前超声心动图示左心室射血分数 (LVEF, $59\% \pm 6\%$), 较术前无明显改善 ($P = 0.10$), LVEDD (46.9 ± 5.9) mm 较术前显著缩小 ($P = 0.02$), 6 例二尖瓣成形患者中, 1 例由术前的重度变为术后的轻度, 5 例中度变为术后微量。

随访 35 例, 平均 5.5 年, 最长 26 年, 随访期间无死亡, 2 例肺动脉内隧道瘘, 1 例行介入封堵, 1 例再次手术修补; 所有患者心功能分级 (NYHA) I 或 II 级; 最后一次随访 LVEF $69\% \pm 7\%$, LVEDD (48.7 ± 5.9) mm; MR 中度 2 例, 轻度 10 例。

3 讨论

3.1 ALCAPA 发病率和病理生理特点

ALCAPA 是一种罕见的先心病, 目前根据临床表现和发病特点分为婴儿型和成人型两类^[4]。90% 的患者早期随着肺动脉压力的逐步下降, 心肌缺血逐渐加重, 1 岁内因心力衰竭死亡, 这类患者为婴儿型。10% 患儿由于左右冠状动脉之间形成良好的侧枝循环而减少左心室心肌缺血, 进而生存至成年, 这类患者为成人型^[5]。

成人患者冠状动脉血流循环方向为右冠状动脉-侧枝循环-左冠状动脉-肺动脉, 这一核心特点决定了患者临床表现、诊断和治疗选择。由于侧枝循环丰富, 心肌缺血不显著, 因此发病隐匿^[6-7], 但是如果不及及时诊治, 也会导致恶性心律失常、猝死可能^[8]。本组患者 29 例患者为活动后而不是静息状态下胸闷或胸痛, 主要原因是所有心肌血供都通过右冠状动脉供应, 因此在心脏负担增加时才导致心肌相对缺血而出现症状。同样由于所有血供来自于右冠状动脉, 右冠状动脉会代偿性的增粗和迂

曲,这与冠状动脉瘘特点类似,这就增加了两者之间相互误诊的概率,对于可疑患者应该进一步行 CT 或造影明确左冠状动脉开口位置以确诊^[9-10]。

3.2 ALCAPA 外科治疗发展历程和结果

ALCAPA 患者的外科治疗经历了以下几个发展阶段。第一个阶段是结扎异常起源的左冠,其主要目的是通过减少冠状动脉系统向肺动脉的分流而间接增加冠状动脉血流灌注,因大部分患者左右冠状动脉之间侧枝循环建立不充分,结扎后冠状动脉血流增加不显著,导致手术的近期和远期死亡率较高,目前这种方法几乎不被采用;第二个阶段是在结扎的基础上,联合左冠状动脉系统的冠状动脉旁路移植,这种手术方式明显增加冠状动脉血流灌注,对于小儿患者而言,更为细小的冠状动脉以及桥血管不仅增加手术难度,而且桥血管远期通畅率远低于成人患者,无法满足小儿患者预期寿命的需求,对于成人患者而言,主要应用于冠状动脉开口细小或者壁内走行患者,本组有 3 例患者采用此法;第三个阶段是肺动脉内隧道修补术(Takeuchi 术)^[11],即先建立主动脉-肺动脉窗口,然后将部分肺动脉血管壁游离,在肺动脉内应用自体部分肺动脉壁建立隧道,将左冠状动脉开口通过内隧道延续至主动脉,恢复了左冠状动脉的正常起源和灌注,但内隧道在肺动脉内有血流的长期冲刷,容易导致肺动脉内隧道瘘或者肺动脉狭窄,目前这种方法主要应用于冠状动脉开口离主动脉较远而无法再植的患者^[12-13]。本组有 16 例患者采用此法。随访过程中 2 例患者出现内隧道漏,并出现明显分流,分别采用介入修补和外科修补,肺动脉瘘的治疗原则和方法与左冠状动脉肺动脉瘘相同,即瘘口>3 mm,因冠状动脉窃血导致了心肌缺血临床表现或者因分流量大导致左心室扩大的表现时应该积极干预,介入封堵对于漏口<8 mm 患者可以尝试,对于漏口较大患者应该手术修补漏口。第四个阶段是冠状动脉再植术^[13-14],即将左冠状动脉开口从肺动脉游离,然后再植入主动脉,这种方法由于生理和解剖上完全恢复左冠系统,因此近期和远期结果良好,已成为首选,但对术者技术要求较高,本组 17 例患者采用此法。

3.3 二尖瓣反流的处理原则

目前对于婴幼儿患者合并二尖瓣反流的处理尚存争议^[15-16]。但成人患者具有不同的特点,因为有良好的侧枝循环,二尖瓣反流发病率及严重程度均低于婴幼儿^[17-18]。本组数据仅 1 例重度反流,5 例中度反流。相对于小儿二尖瓣成形而言,成人二尖

瓣成形技术普及程度广、难度相对低、效果确切,因此对于成人 ALCAPA 患者并发二尖瓣反流的处理可以遵守单纯二尖瓣反流的治疗原则,即对于中度以上反流应积极修复;由于该类患者二尖瓣反流多为瓣环扩大导致,因此瓣环成形是最常用和有效的方法,还需要考虑部分患者乳头肌缺血坏死纤维化,必要时应该行乳头肌松解,以增加瓣叶活动度。本组数据显示二尖瓣成形近期和远期效果满意。

综上所述,成人左冠状动脉起源于肺动脉发病率极低,诊断明确即应手术治疗,肺动脉内隧道术可能由于远期内瘘形成需要再次干预,对于合并中度以上二尖瓣反流患者,同期行二尖瓣成形术具有良好效果。

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