

• 研究报告 •

先天性冠状动脉瘘早中期手术疗效分析*

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[摘要] **目的:**探讨先天性冠状动脉瘘手术治疗的时机、方法及疗效,努力提高手术成功率。**方法:**我院2013年10月—2019年10月共收治19例冠状动脉瘘患者,其中男11例,女8例,年龄4.5(1~61)岁,体重16.25(7~69) kg。右冠状动脉右房瘘3例,右冠状动脉右室瘘7例,右冠状动脉左室瘘1例,左冠状动脉右房瘘2例,左冠状动脉左房瘘2例,左冠状动脉右室瘘2例,左冠状动脉肺动脉瘘2例;合并先天性二尖瓣发育不良2例,房间隔缺损、室间隔缺损、卵圆孔未闭、三尖瓣关闭不全各1例。所有患者均胸正中切口体外循环下行动脉瘘矫治术,同期矫治其他心脏畸形,围术期常规监护治疗,术后1、6个月及每年随访复查。对所有患者临床资料进行统计、分析及总结。**结果:**升主动脉阻断时间(61.330±11.479) min,体外循环时间(99.940±15.206) min,手术时间(140.610±19.150) min,呼吸机使用时间(33.000±19.275) h,ICU时间(2.220±1.215) d,住院时间(20.940±8.795) d。患者术前左室射血分数(63.167±8.031)%,术后当日左室射血分数(69.390±8.389)%,术后左室功能明显改善($P<0.05$)。患者术前心胸比0.506±0.0769,术后当日心胸比0.479±0.0603,手术前后心胸比差异显著($P<0.05$)。19例患者围手术期无严重并发症及死亡,均痊愈出院。术后随访6个月~5年,未见残余瘘及严重心血管症状。**结论:**冠状动脉瘘发病率低,出现心悸、呼吸困难等症状应尽早手术治疗,外科手术疗效较好,并发症少。

[关键词] 先天性冠状动脉瘘;外科治疗;预后

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Short and medium-term surgical treatment effects of congenital coronary artery fistula

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Abstract Objective: To summarize clinical experiences and surgical outcomes of congenital coronary arterial fistula (CAF), and to provide evidence for surgical decision and postoperative recovery and improving the success rate of surgery. **Method:** We retrospectively analyzed clinical records of 19 patients who underwent surgical repair of CAF in our center from October 2013 to October 2019, including 11 males and 8 females. The median age was 4.5(1-61) years, and the median weight was 16.25(7-69) kg. There were 3 cases with right coronary artery-right atrial fistula, 7 cases with right coronary artery-right ventricular fistula, 1 case with right coronary artery-left ventricular fistula, 2 cases with left coronary artery-right atrial fistula, 2 cases with left coronary artery-left atrial fistula, 2 cases with left coronary artery-right ventricular fistula, and 2 cases with anomalous left coronary artery from the pulmonary artery. Two patients complicated with congenital mitral valve dysplasia, 1 case complicated with atrial septal defect, 1 case complicated with ventricular septal defect, 1 case complicated with patent foramen ovale, and 1 case complicated with tricuspid insufficiency. All patients underwent CAF correction under cardiopulmonary bypass through thoracic median incision, and other cardiac malformations were corrected at the same time. Routine monitoring and treatment were performed in the perioperative period, 1 month, 6 months, and annual follow-up after operation were conducted. The clinical data of all patients were statistically analyzed and summarized. **Result:** The aortic cross-clamping time was (61.33±11.479) min, the cardiopulmonary bypass time was (99.94±15.206) min, the operation time was (140.61±19.150) min, the intubation duration was (33±19.275) h, the length of ICU

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stay was (2.22±1.215) d, and the length of hospital stay was (20.94±8.795) d. The preoperative left ventricular ejection fraction (LVEF) was (63.167±8.031)%, the postoperative LVEF was (69.39±8.389)%, and the postoperative left ventricular function were significantly improved ($P<0.05$). The preoperative cardiothoracic ratio was 0.506±0.0769, and the postoperative cardiothoracic ratio was 0.479±0.0603, and the difference was statistically significant ($P=0.006<0.05$). No death was found during the perioperative period, all patients were recovered and discharged. Followed up for 6 months to 5 years, no severe cardiovascular symptoms related to residual fistula was found. **Conclusion:** The incidence of CAF is low. A few patients can appear palpitations, breathing difficulties and other symptoms, which should be operated as soon as possible. Surgical treatment is effective and has less complications.

Key words coronary artery fistula; surgical treatment; prognosis

冠状动脉瘘(coronary artery fistula, CAF)是冠状动脉(冠脉)与心腔或大血管之间的异常连接,是一种罕见的先天性缺陷,约占总人群的0.002%,占有冠脉异常的14%^[1]。其临床相关性主要集中在“冠脉窃血现象”的机制上,即使没有狭窄,也可引起心肌功能缺血,常见症状为心绞痛或呼吸困难。如果无明确症状则可进行临床随访,对于有症状的、较大的或巨大的瘘管,经导管入路或手术结扎的侵入性治疗是合理的治疗手段。

1 对象与方法

1.1 对象

本中心2013年10月—2019年10月诊治CAF患者19例,其中男11例,女8例,年龄4.5(1~61)岁,体重16.25(7~69) kg。患者术前一般资料见表1。

表1 患者基本临床资料

Table 1 General information of included patients 例

指标	例数
瘘类型	
右冠-右房	3
右冠-右室	7
右冠-左室	1
左冠-右房	2
左冠-左房	2
左冠-右室	2
左冠-肺动脉	2
术前症状	
有症状(心悸、气促等)	15
无症状	4
心电图	
ST段改变	3
电轴左偏	1
窦性心动过速	1
正常心电图	14
合并畸形	
先天性二尖瓣发育不良	2
房间隔缺损	1
室间隔缺损	1
卵圆孔未闭	1
三尖瓣关闭不全	1
右室异常肌束	1

1.2 术前评估

1.2.1 超声心动图 超声心动图是非侵入性的,是评价心室大小和功能、冠脉扩张及其程度、瘘口流速、瓣膜反流程度和肺动脉压的有效方法。另外,对于多发瘘口、小瘘口和巨大瘘口患者,术中行经食管超声心动图检查,可以更加精确地诊断,并对术后是否有残余分流做出及时判断^[2]。

1.2.2 CTA 在严重患者影像学术前评估中,CT优于超声心动图。多排CT具有很好的时间和空间分辨率,再结合心电门控、图像后处理等技术,能清楚地显示冠脉的起源、走行、瘘口部位、数量及“窃血情况”,更好地指导手术方式的选择^[3-4]。本研究各种类型冠脉瘘CTA图像见图1。

1.3 手术适应证及手术方法

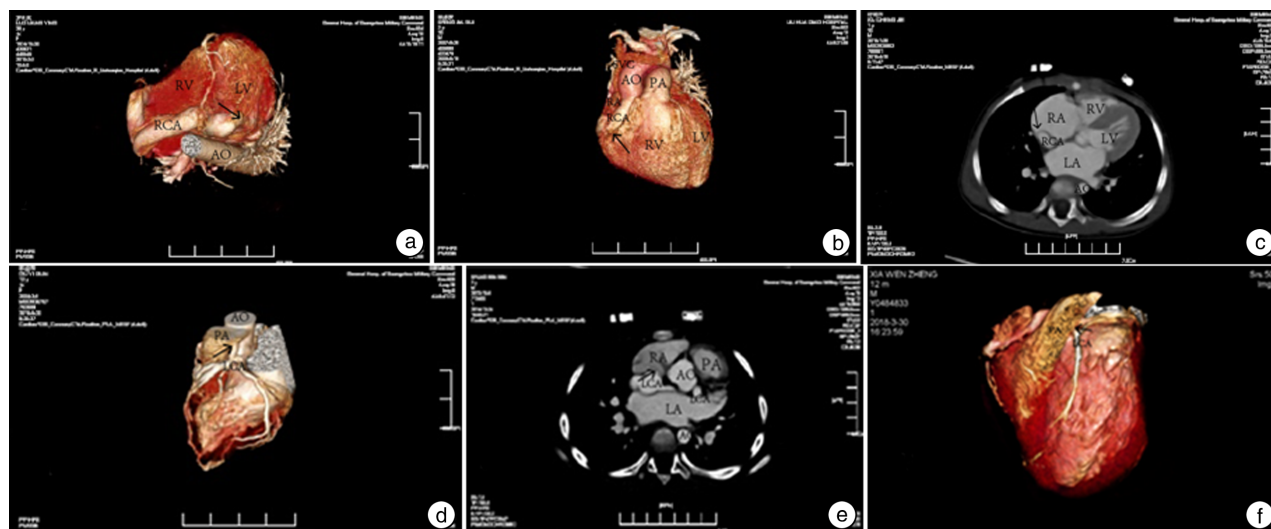
适应证包括:分流量较大、合并有其他心脏畸形、冠脉迂曲膨大明显、伴有冠脉瘤形成、急性成角、解剖复杂、危及邻近血管的CAF。手术正中胸骨切口,心外探查CAF,探查瘘管走行。常规建立体外循环,体温降至32℃时,阻断上、下腔静脉、升主动脉,冷灌心脏停跳(适当夹闭扩张冠脉避免停跳液分流),根据瘘的类型选择切开心房或心室或直接缝合瘘管以关闭瘘管的入口(冠脉端)和出口(心房、心室、腔静脉、肺动脉端),若伴有冠脉瘤,平行冠脉走行方向切开冠脉瘤予以成形;合并畸形行一期修复。开放升主动脉,心脏复跳后常规停循环、关胸。本组CAF患者合并冠脉瘤4例,3例行CAF矫治+冠脉瘤成形术,1例行CAF矫治术+冠脉瘤切除术+冠脉旁路移植术。1例合并房间隔缺损行修补术,1例合并房间隔缺损修补术,1例合并卵圆孔未闭缝合,2例先天性二尖瓣发育不全及1例三尖瓣关闭不全同期行二尖瓣成形及三尖瓣成形术。

1.4 术后评估及随访

所有患者术后当日行心脏超声检查、胸片检查及心电监护,评估有无残余瘘、心功能情况、心胸比值及心律情况。患者采用门诊复诊方式随访,复查心电图、心脏超声了解康复情况。

1.5 统计学处理

应用SPSS 20.0进行数据处理。正态分布定



a:右冠脉左室瘘,右冠脉显著扩张且从主动脉根部发出,汇入左室;b:右冠右室瘘,右冠脉从主动脉根部发出,汇入右室;c:右冠脉右房瘘,右冠脉显著扩张从主动脉根部发出,汇入右房;e:左冠脉右房瘘,左冠脉从主动脉根部发出,汇入右房,且在入右房处呈动脉瘤扩张,压迫右房;d,f:左冠脉肺动脉瘘,左冠脉从肺动脉主干后壁发出。各图中箭头指向瘘管汇入位置。AO:主动脉;LA:左房;LV:左室;RA:右房;RV:右室;RCA:右冠脉;LCA:左冠脉;SVC:上腔静脉;PA:肺动脉。

图1 典型CAF的CT图

Figure 1 CT images of typical CAF

量资料用 $\bar{x} \pm s$ 表示,非正态分布定量资料用中位数(最小值,最大值)[M(min, max)]表示,术前、术后和随访射血分数、心胸比比较采用配对样本 t 检验,术前、术后和随访心功能分级比较采用 χ^2 检验, $P < 0.05$ 为差异有统计学意义。

2 结果

2.1 术中及术后早期结果

19例患者均行CAF矫治术,手术时间(140.610 ± 19.150) min,升主动脉阻断时间(61.330 ± 11.479) min,体外循环时间(99.940 ± 15.206) min,呼吸机时间(33.000 ± 19.275) h,ICU时间(2.220 ± 1.215) d,住院时间(20.940 ± 8.795) d。患者术前左室射血分数(63.167 ± 8.031)%,术后左室射血分数(69.390 ± 8.389)%,与术前相比,术后左室功能明显改善($P < 0.05$)。患者术前心胸比 0.506 ± 0.0769 ,术后心胸比 0.479 ± 0.0603 ,手术前后心胸比差异有统计学意义($P < 0.05$)。术后心功能分级与术前相比差异无统计学意义。

2.2 术后并发症及死亡

患者术后循环及内环境稳定,无严重并发症及死亡,查心脏超声未见残余瘘,心电图检查未出现严重心律失常等异常情况,患者症状得到改善,均痊愈出院。

2.3 随访结果

19例患者随访6个月~5年。随访期间,患者左室射血分数与术前及术后相比差异均有统计学意义(均 $P < 0.05$),左心功能有所改善;患者心胸比值与术前及术后相比差异均有统计学意义(均 $P < 0.05$),心脏容量过载现象得到改善;患者心功能分级与术前及术后比较均差异无统计学意义;患者心电图检查无明显异常;心脏彩超显示心功能均可,未发现残余瘘,无因心脏事件再次手术的患者。详见表2。

3 讨论

CAF是Krause在1865年定义的一类冠脉终点异常畸形,占先天性心脏病的 $0.27\% \sim 0.4\%$ ^[5]。80%的CAF是单纯性的,但也可合并其他心脏畸

表2 患者术前、术后及随访数据比较

Table 2 Preoperative, postoperative and followed-up data of patients

阶段	左室射血分数/%	纽约心功能分级/例(%)				心胸比值
		I	II	III	IV	
术前	63.167 ± 8.031	8(42.0)	7(36.8)	3(15.8)	1(5.3)	0.506 ± 0.0769
术后	69.390 ± 8.389 ¹⁾	13(68.4)	5(26.3)	1(5.3)	0(0)	0.479 ± 0.0603 ¹⁾
随访	71.000 ± 6.954 ^{1) 2)}	15(78.9)	3(15.8)	1(5.3)	0(0)	0.449 ± 0.405 ^{1) 2)}

与术前比较,¹⁾ $P < 0.05$;与术后比较,²⁾ $P < 0.05$ 。

形,约占20%,包括房间隔缺损、动脉导管未闭、室间隔缺损、肺动脉闭锁、法洛三联症等^[6]。本研究患者合并室间隔缺损、房间隔缺损、卵圆孔未闭各1例,均行同期修复。

本研究术后及随访左心射血分数对比术前有显著改善,显示手术消除了“冠脉窃血现象”,患者心功能得到明显改善。研究表明,严重的“冠脉窃血现象”,可导致心肌缺血,出现可逆或不可逆的心肌功能障碍、静息性心绞痛等症状,其严重程度与“窃血现象”、瘘管分流量大小和血管之间的压力阶差密切相关^[7-8]。所以术前影像学准确地评估患者病情异常重要。

本研究患者经手术治疗后,术后及随访过程中,心脏容量过载现象得到缓解。CAF患者心脏容量过载的原因主要是瘘管的存在导致冠脉血流流向左心或右心系统,使左心或右心出现容量过载现象,可能导致心力衰竭、心律失常等症状^[9]。患者经手术封闭瘘管后,终止了异常分流的现象,使得心脏容量过载得到缓解,影像学上表现为心脏有所缩小,心胸比值减小。本研究术后、随访与术前数据对比结果也证实了这一点。

CAF中高血流可导致冠脉扩张和动脉瘤,同时这种异常的血流也可导致乳头肌功能障碍,从而出现瓣膜反流的症状^[10]。本研究对发现冠脉瘤的4例患者及瓣膜反流的3例患者均行手术治疗。4例冠脉瘤患者中,3例行冠脉瘤切开,切除部分组织后重新缝合,1例左冠脉右房瘘因瘤体较大压迫右房,行冠脉瘤切除+冠脉旁路移植术。3例瓣膜反流患者行同期瓣膜成形术。CAF可以起源于任何冠脉的主干,但以右冠脉较多。本组CAF患者中源于右冠脉12例,源于左冠脉7例,与报道较为相符。同起始冠脉一样,CAF的汇入位置理论上可以是任意心腔和血管,但以腔内压较低的右心房、右心室、肺动脉多见^[11-12]。本组19例病例中,汇入右室9例,汇入右房5例,汇入肺动脉1例,与文献报道比例基本相符^[13]。

对于手术时机,美国心脏病学会指南指出,出现严重的“冠脉窃血”和心血管症状的小、中型瘘成年患者,应结扎瘘管,避免出现心肌缺血、心律失常、心室功能障碍及瘘相关心内膜炎等症状的风险;对于无症状瘘管较小的成年患者,应通过每3~5年1次的超声心动图进行临床随访管理^[14]。对于儿童,尤其是对于5岁以上的儿童,即使无症状,也应选择关闭瘘管较大的瘘道^[15-16]。此外,有研究强烈建议关闭发生在冠脉近段的瘘管,因为这种类型的瘘管更有可能变成动脉瘤,并有较高的破裂风险^[17]。本研究中,成人患者5例,均因出现心悸、气促等症状入院,4例为中型瘘管,1例为大型瘘管,直径为10 mm,呈瘤状。有报道指出,对于直径

>8 mm的巨大动脉瘤,有更高的风险发展为狭窄,预后更差。建议使用阿司匹林进行药物治疗,应至少服用6~8周;对于巨大动脉瘤,无论是否使用华法林或低分子量肝素,都应使用抗血小板药物(阿司匹林和氯吡格雷),同时使用 β 受体阻滞剂用于降低心肌耗氧量^[18]。余14例为儿童患者,1~5岁的有8例,5岁以上的有6例,其中10例出现气促、杂音等症状,4例未出现明显症状。所有患者均按指南的手术时机及指征行手术治疗,合并室间隔缺损、房间隔缺损和卵圆孔未闭等畸形行同期修补、缝合。

患者术后循环及内环境稳定,术后复查无血栓形成、出血、心肌缺血、感染性心内膜炎和严重心律失常等并发症,查心脏超声未见残余瘘,住院期间无死亡。但术后有3例出现左室收缩功能降低,射血分数下降,均为儿童,其中2例为右冠脉左心室瘘,1例为左冠脉肺动脉瘘。由于儿童冠脉较细小,在使用自体心包片修补瘘口时,切开和缝合冠脉时有可能损伤冠脉分支,射血分数的下降可能与此有关。随访过程中,未发现残余瘘,症状得到较大改善,无因心脏事件再次手术的患者。仅有1例老年患者因心功能不全再次住院,行保守治疗病情稳定后出院。CAF大部分是体检发现心脏杂音或合并其他疾病进行超声心动图和CTA检查时发现。最近Uchida等^[19]报道了一种术中荧光成像的方法诊治CAF,较新颖,有助于术中更加清晰地了解CAF的走向。外科手术闭合瘘道能治疗95%以上的CAF,常用的方法有瘘管结扎术、经心腔瘘口关闭术和冠脉切开修补术等^[8,20]。当然,随着手术技术的发展,一些中心也在探索新的手术方法,如经导管闭合、全胸腔镜下闭合、3D打印模型指导闭合和运用虚拟现实技术闭合等^[21-26]。其中又以经导管闭合CAF应用较多,但是也会出现与术式相关的并发症,如穿刺部位血肿形成、血管夹层或穿孔、瓣膜损伤、封堵装置术后移位等,需要严格掌握适应证。本单位采用传统的开胸闭合,术后及随访效果较好,症状得到改善,且未发现残余漏。但也有研究认为CAF的治疗不能盲目追求手术的微创性,应根据患儿具体病情制定合适的手术方案。

总之,CAF发病率较低,良好的影像学资料对手术异常重要,对有手术指征的患者进行早期手术治疗的效果良好,术者应根据患者的情况选择合适的术式。

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