

肥厚梗阻性心肌病的外科治疗*

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摘要 目的:总结肥厚室间隔切除术治疗肥厚梗阻性心肌病的手术效果,探讨外科治疗策略。方法:2002年3月至2010年10月,外科手术治疗33例肥厚梗阻性心肌病患者。其中男16例,女17例;年龄13~59岁,平均(42.7±13.6)岁;左室流出道压差(LVOTGP)70~120 mmHg(1 mmHg=0.133Kpa),平均(95.0±22.6)mmHg。其中合并二尖瓣关闭不全24例,主动脉瓣关闭不全7例,升主动脉增宽3例,冠心病2例。手术在全麻低温体外循环下完成,按常规经主动脉切口行室间隔心肌切除术,同期完成二尖瓣置换术(MVR)7例,二尖瓣成形术(MVP)7例,二尖瓣、主动脉瓣成形术(MVP+AVP)5例,二尖瓣、升主动脉成形术3例,二尖瓣、主动脉瓣成形、冠状动脉旁路移植术(MVP+AVP+CABG)2例。分析比较病人术前超声心动图(UCG),术中经食管心脏超声(TEE),以及术后1周、3月、6月、1年超声心动图结果。结果:手术死亡1例(3.0%,1/33例),主要死因为严重低心排综合症以及多脏器功能衰竭。二次开胸止血1例(3.0%,1/33例)。术中经食管心脏超声示所有病人二尖瓣前叶收缩期前向运动现象(SAM征)消失。存活病人手术效果良好,解剖狭窄解除,峰值压差降低,SAM现象基本消失。远期随访生存病人症状消失,生活质量明显改善,心功能I~II级,无远期死亡、并发症或再次手术。结论:外科治疗肥厚梗阻性心肌病具有良好的手术效果。了解病理生理过程、术中仔细探查、手术切除彻底是手术成功的关键。

关键词 心肌病 肥厚性 心脏外科手 SAM征

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Surgical Treatment of Hypertrophic Obstructive Cardiomyopathy*

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ABSTRACT Objective: To summarize the results of hypertrophic ventricular septum myectomy for hypertrophic obstructive cardiomyopathy (HOCM), and investigate surgical treatment strategies. **Methods:** From March 2002 to October 2010, 33 patients of HOCM underwent surgical treatment. There were 16 males and 17 female patients. The age ranged from 13 to 59 years old with a mean of (42.7±13.6) years old. The left ventricular out tract gradient pressure (LVOTGP) was 70~120 mmHg (1 mmHg = 0.133Kpa), with a mean of (95.0±22.6) mmHg. 24 patients combine mitral valve regurgitation, 7 patients combine aortic valve insufficiency, and 3 patients with ascending aortic widened 2 patients with coronary heart disease. Surgical operations accomplished under hypothermic cardiopulmonary bypass with general anesthesia, and ventricular septum myectomy carried through conventional aortic incision. There were 7 patients with mitral valve replacement (MVR), 7 patients with mitral valvuloplasty (MVP), 5 patients with mitral valve aortic valvuloplasty (MVP + AVP), 3 patients with mitral ascending aorta valvuloplasty, and 2 patients with mitral valve aortic valvuloplasty, coronary artery bypass grafting over the same period (MVP + AVP + CABG). The manifestation of pre-operative UCG, intro-operative transesophageal echocardiography (TEE) and post-operative UCG in 1 week, 3 months, 6 months, 1 year were compared and analyzed. **Results:** 1 patient (3.0%, 1 / 33) died in hospital due to severe low cardiac output syndrome and multiple organ failure. 1 patient (3.0%, 1 / 33) carried Second thoracotomy to stop bleeding. Intro-operative TEE showed systolic anterior mitral leaflet in all patients prior to movement phenomena (SAM sign) disappear. Survival of patients with good effect of surgery, anatomical narrow lift, lower peak pressure, SAM phenomenon disappeared. Long-term survival of the patients were followed up for symptoms, quality of life significantly improved cardiac function in I~II class, no late death, complications or reoperation. **Conclusion:** Surgical treatment of hypertrophic obstructive cardiomyopathy is safe and efficient. Understand the pathophysiological process, careful exploratory surgery, radical surgery is the key to successful operation.

Key words Cardiomyopathy; hypertrophic; Cardiac surgical procedures; Systolic anterior movement

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前言

肥厚梗阻性心肌病(HOCM)是一种常染色体显性遗传为主,室间隔和左心室壁心肌肥厚使心肌收缩力增强,舒张期顺应性下降的原发性心肌病。近年来,随着其病理生理的进一步认识,心血管外科诊疗技术的成熟和发展,室间隔肥厚肌束切除术治疗肥厚梗阻性心肌病取得了显著进步,是缓解左室流出道梗阻最可靠的治疗方法。HOCM常常合并不同程度的二尖瓣反流,部分病人同时合并主动脉以及冠状动脉病变,故临床治疗中往往需要在缓解左室流出道梗阻的同时,纠正二尖瓣关闭不全等其他心脏病变。

2002年3月至2010年10月,我院为33例肥厚梗阻性心肌病病人行室间隔肥厚肌束切除术,现结合本组治疗经验分析肥厚梗阻性心肌病的病理生理特点,对其疗效进行分析并探讨其外科治疗策略。

1 资料和方法

1.1 一般资料

本组患者共33例,其中男16例,女17例,年龄13~59岁,平均(42.7±13.6)岁。主要症状为胸闷、心悸、心前区疼痛、晕厥等,大部分病人服用β受体阻滞剂和钙通道拮抗剂,因症状不能控制而选择手术治疗。查体示胸骨左缘3~4肋间粗糙收缩期杂音伴有震颤。术前心电图示左室肥厚,X线胸片示心脏轻度增大。术前超声心动图示左室壁及室间隔非对称性肥厚,典型的二尖瓣前叶收缩期前向运动(SAM征),左室流出道压差(LVOTPG)70~120 mmHg,平均(95.0±22.6)mmHg。其中合并二尖瓣关闭不全24例,主动脉瓣关闭不全7例,升主动脉增宽3例,冠心病2例(表1)。

1.2 手术方法

手术均在全身麻醉低温体外循环下进行,术中常规行食管心脏超声(TEE)监测。常规升主动脉及上、下腔静脉插管建立体外循环。心脏停跳后行升主动脉切口,牵开主动脉瓣叶,暴露

凸向左室流出道前壁的肥厚室间隔(图1),在左室流出道中垫上湿小纱布,自右冠瓣中点右方1~3mm,平行于左室流出道长轴做一纵行切口,在左右半月瓣交界再做一纵行切口,再在两者之间做一横切口,切除肥厚室间隔心肌组织(图2)。切割深度应控制在15~20mm,避免切穿室间隔和损伤传导束。为消除SAM征,切口尽量向左室心尖方向延伸,必要时还切除部分左室游离壁肥厚肌肉,最大限度地扩大左室流出道。对合并二尖瓣关闭不全、主动脉病变及冠状动脉粥样硬化性心脏病的病人,同期行二尖瓣置换或成形术、主动脉成形术及冠状动脉旁路移植术。心脏复跳后,常规TEE检测左室流出道压力及室间隔厚度。

1.3 统计方法

定量资料用均数±标准差($\bar{x} \pm s$)表示,采用SPSS13.0统计学软件进行分析,组内两样本均数采用配对比较t检验, $P < 0.05$ 为差异有统计学意义。

2 结果

全组体外循环68~144min,平均(96.2±31.6)min,主动脉阻断48~106min,平均(68.3±37.5)min,呼吸机辅助时间7~18h,平均(11.6±4.6)h,ICU住院时间18~126h,平均(48.4±26.7)h。

全组死亡1例,病死率3.0%(1/33例),为严术后重低心排综合征以及多脏器功能衰竭。二次开胸止血1例(3.0%,1/33例)。心脏复跳后,TEE显示SAM征均消失。32例获得11~84个月的随访,平均(35±12)个月,生存病人症状消失,生活质量明显改善,心功能I~II级,无远期死亡、并发症或再次手术,随访中进行超声心动图检查。术中TEE,术后1周、3个月、6个月及1年的超声心动图均提示左房内径(LAD)、左室舒张末径(LVDd)、左室流出道压差(LVOTPG)、室间隔厚度(IVST)较术前显著下降($P < 0.05$);除术前外,其他时间各点的LAD、LVDd、LVOTPG、IVST间差异无统计学意义(表2)。

表1 病人基本资料

Table 1 Basic information of patients

手术方式 (surgical method)	例数	男性	有 HOCM 家族史 (例 %)	有黑朦或晕厥史 (例 %)	术前心功能 III/IV 级 (NYHA) 例
Simple excision	9	6	3 33.3%	4 44.4%	3
Simple excision +MVR	7	3	1 14.3%	2 28.6%	1
Simple excision +MVP	7	2	2 28.6%	3 42.9%	2
Simple excision +AVP+MVP	5	3	0	2 40.0%	1
Simple excision +MVP+ascending aorta plasty	3	1	0	1 33.3%	1
Simple excision +CABG+MVP+AVP	2	1	0	1 50%	

3 讨论

HOCM 是肥厚性心肌病中比较常见的一种类型,又称特

发肥厚性主动脉瓣下狭窄,主要因室间隔肥厚导致左室流出道梗阻得名^[1]。此类肥厚性心肌病发病率约为0.2%^[2],可出现于任何年龄,20~40岁多见。约1/3肥厚性心肌病病人有明确家族

史^[3],常伴有二尖瓣前叶收缩期前向运动现象(SAM 征),非对称性室间隔肥厚和 SAM 征是其病理特点^[4]。左室流出道梗阻造成收缩期压力阶差,梗阻期"射流效应"引起的 SAM 征进一步加重了左室流出道的梗阻,进而室壁顺应性下降,心肌代偿性肥厚,心肌缺血及心律失常,甚至突发心源性死亡。国外关于肥厚性心肌病人长期预后的研究显示,1/3 年轻运动员猝死是由于心肌病^[5],其年病死率为 1.4%~2.2%,5 年生存率为 89%~93%^[6-8]。

表 2 病人术前、术中和术后超声心动图结果比较($\bar{x} \pm s$)
Table 2 Comparison of preoperative, intraoperative, and postoperative echocardiography

时间 Time	LAD (mm)	LVDd (mm)	LVOTPG (mmHg)	IVST (mm)	LVPWTd (mm)	EF
术前 preoperative	43.7± 7.2	45.4± 5.1	95.0± 22.6	20.5± 4.2	14.4± 3.8	0.69± 0.05
术中 TEE intraoperative TEE	37.2± 6.5a	41.8± 6.4	22.8± 8.7a	13.6± 2.3a	13.8± 4.2	0.64± 0.09
术后 1 周 Postoperative 1 week	35.6± 5.4a	37.6± 7.1	21.9± 7.9a	13.4± 3.8a	13.6± 5.7	0.63± 0.04
术后 3 月 Postoperative 3 months	34.2± 5.8a	36.8± 6.3	20.4± 8.2a	13.8± 4.2a	12.8± 4.9	0.61± 0.07
术后 6 月 Postoperative 6 months	33.8± 6.2a	36.4± 6.1	21.6± 5.8a	12.7± 5.6a	12.3± 5.2	0.63± 0.06
术后 1 年 Postoperative 1 year	34.7± 7.4a	35.7± 5.2	21.2± 6.4a	12.4± 6.1a	12.6± 4.8	0.61± 0.04

a 与术前相比 P<0.05
aCompared with the preoperative P<0.05

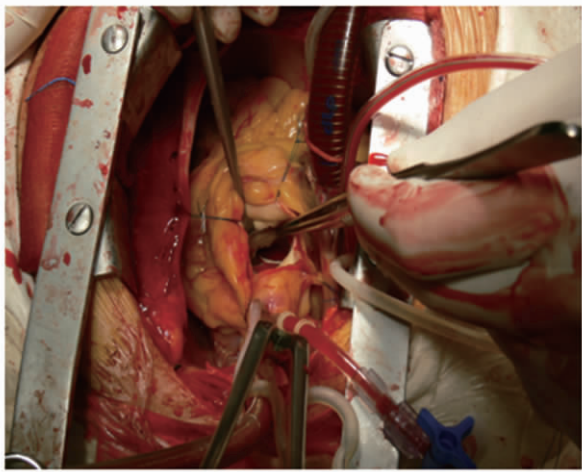


图 1 主动脉瓣下肥厚的室间隔
Fig.1 hypertrophic ventricular septum under Aortic valve



图 2 切除的室间隔心肌组织
Fig.2 Resection of the ventricular septum myocardium

HOCM 病人治疗根据临床症状轻重及左室流出道压力阶差有多种方法。对于跨瓣压差 <30mmHg 的无症状病人,可以口服 β 受体阻滞剂和钙通道拮抗剂等药物控制,也可通过双腔起搏器植入和经皮室间隔心肌化学消融术等手段治疗。但对于上述治疗效果不满意的中、重度病人,或药物激发试验跨瓣压差 >50mmHg 的病颤病人,跨瓣压差 >100mmHg 无临床症状的病人,有明确猝死家族史的病人都都是外科手术指针^[9]。目前,室间隔心肌切除术仍是治疗 HOCM 的金标准^[10]。

HOCM 常伴有二尖瓣关闭不全,二尖瓣自身病变占病人总数的 4%~14%^[11,12]。病变主要包括瓣膜炎、异常的乳头肌或

腱索、粘液样变性和退行性变性等^[13],同期需要处理二尖瓣问题,大部分二尖瓣成形术后可取的满意效果,但也有 10%~20%的病人二尖瓣本身有先天性发育异常或激发性病变,可予以置换^[14]。Ryan 等^[15]报道,约 58%的病人二尖瓣可以得到很好的修复,由于人工瓣膜不可避免地带来感染、抗凝并发症等风险,以及外科瓣膜成形技术的进步,成形术在相当病例中越来越被外科医生所运用。本组共有 24 例病人不同程度的伴有二尖瓣关闭不全,17 例予以成形术,7 例予以置换术,术中 TEE 显示效果满意。单纯室间隔心肌切除术手术病死率文献报道 <1%^[16],如同期行冠状动脉旁路移植术或升主动脉成形、主动

脉瓣成形术,手术病死率 <5%^[17]。本组同期合并升主动脉成形 3 例,冠状动脉旁路移植 2 例,主动脉瓣成形 7 例,手术效果满意,仅有 1 例因术后严重低心排综合症以及多脏器功能衰竭死亡,生存病人远期随访症状消失,生活质量明显改善。

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