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法洛四联症外科治疗

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摘　要

法洛四联症是最常见的先天性心脏病之一，位居紫绀型先天性心脏病的首位，右心室流出道严重梗阻的婴儿未进行手术干预的死亡率为25%，3岁时死亡率为40%，10岁时死亡率为70%，40岁时死亡率为95%，因此尽早进行外科干预十分必要。现参考国内外近年来相关文献，对法洛四联症姑息手术、根治手术的发展历史，一些面临的主要问题及主要并发症做一综述。

关键词

法洛四联症，姑息治疗，根治术，并发症

Surgical Treatment of Tetralogy of Fallot

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Abstract

Tetralogy of Fallot is one of the most common congenital heart diseases, ranking first in cyanotic congenital heart disease. The mortality rate of infants with severe right ventricular outflow tract obstruction without surgical intervention was 25%, 40% at 3 years old, 70% at 10 years old and 95% at 40 years old, so it is necessary to carry out surgical intervention as soon as possible. Referring to the relevant literature at home and abroad in recent years, this paper reviews the development history, main problems and complications of palliative and radical surgery for tetralogy of Fallot.

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Keywords

Tetralogy of Fallot, Palliative Treatment, Radical Surgery, Complications

1. 引言

法洛四联症(tetralogy of Fallot, TOF)是最常见的紫绀型先天性心脏病，患病率约为全部先天性心脏病的 12%~14%，为紫绀型先天性心脏病的 50%~90%，新生儿中发病率为 0.034% [1]。该病在 1671 年由丹麦医生 Niels Stenson 首次报道，在一篇题为《巴黎怪胎解剖》的短文中描述了动脉出现的不寻常形式，肺动脉狭窄，动脉导管缺失，主动脉下室间隔缺损，双侧心室共同的主动脉管。1777 年荷兰医生 Eduard Sandifort 报告了一例 16 个月大的病人患上了四联症，尸检发现他的心脏先天畸形，没有动脉导管或动脉韧带的迹象。1888 年 Fallot 以详细的四个主要特征描述了该病与其他青紫型心脏疾病的区别，强调紫绀不是由未闭的卵圆孔引起，而是由于宫腔内的病理过程，并表明四联症本质上只是累及肺动脉和漏斗部的一种异常，导致肺狭窄、室间交通、主动脉双室起源和右室肥厚，Fallot 使用的名称是“蓝色疾病”或“发绀心脏病”[2]。1924 年加拿大医生 Maude Elizabeth 正式将右心室流出道(right ventricular outflow tract, RVOT)狭窄、室间隔缺损(ventricular septal defect, VSD)、主动脉起源位置右移和右心室肥厚命名为 TOF。在右室流出道严重梗阻的婴儿出生后一年内，未进行手术干预的死亡率为 25%，3 岁时死亡率为 40%，10 岁时死亡率为 70%，40 岁时死亡率为 95% [3]。在没有手术治疗的患者中，最常见的死亡原因包括缺氧(68%)、脑血管意外(17%)和脑脓肿(13%)，因此尽早对 TOF 患者进行外科干预十分必要[4]。现对 TOF 姑息手术及根治手术的发展历史及面临的一些主要问题及主要并发症做一综述。

2. 外科治疗

2.1. 姑息手术

正常的肺血流在新生儿期对促进肺血管发育和肺泡生成方面起着重要作用[5]。在复杂先天性心脏病手术治疗中，常常由于患儿术前病情严重，肺动脉重度发育不良，不能耐受期根治手术，因此需先行姑息性手术来增加肺的血流。姑息手术可以改善患儿临床症状，促进肺血管的发育，为根治手术创造条件。姑息手术指征主要为伴有严重肺动脉分支发育不良(即 McGoon 比值 < 1.2、Nakata 指数(150 mm^2/m^2) [6]。Nakata 指数是左、右肺动脉近第一分叉处截面积之和与体表面积的比值，正常值 ≥ 330 mm^2/m^2。McGoon 比值是两侧肺动脉直径之和与膈肌水平降主动脉直径的比值，正常值 > 2.0。目前临床上较多采用的增加肺血流的姑息手术包括改良 B-T 分流术(modified Blalock-Taussig shunt, MBT)，中央分流术(Central Shunt, CS)，改良 Waterston 手术等。

TOF 第一例外科治疗是在 1944 年由 Helen Taussig 与 Alfred Blalock 共同完成的[7]。Taussig 观察到，如果动脉导管保持开放，TOF 患者的紫绀症状能得到明显的缓解，为后续的根治手术创造条件。1944 年，Blalock 在 Thomas 的陪伴下，经左前外侧胸部切口为一名 15 个月、体重 4 公斤的 TOF 患儿成功实施了左锁骨下动脉近端与左肺动脉端侧吻合术，术后这名患儿的紫绀症状得到有效的缓解。这种姑息性的治疗手段被命名为 Blalock-Taussig (B-T)分流术，这标志着先天性心脏手术时代的开始。由于 B-T 手术牺牲

了右锁骨下动脉、吻合口易于形成血栓堵塞血管、术侧上肢血管供血不足、较难拆除等原因，远期预后较差[8]。在聚四氟乙烯(poly-tetrafluoroethylene, PTFE)制成的人工管道问世后，1962 年 Klinner，Pasini，Schaudig 等人、1978 年 Stark de Leval 等人先后利用 PTFE 管道将锁骨下动脉与肺动脉连接起来，即 MBT，这种方法不需要牺牲锁骨下动脉、可以选择不同直径的 PTFE 管道来适应生长发育的需要、易于拆除、不易堵塞管道且不易引起肺动脉扭曲及充血性心衰，在一项平均随访 45 个月的研究中显示 MBT 术后左、右、主肺动脉和肺环直径得到了均匀且显著的增长[9]。1946 年 Potts 对 B-T 分流术进行改进，Potts 利用他自己发明的主动脉侧壁阻断钳，借助侧壁阻断技术完成了降主动脉－肺动脉吻合术，即 Potts-Smith (Potts)分流术。Potts 分流术吻合口较大，可充分保持肺动脉供血和全身供氧，但术者往往难以精确把握吻合口直径，吻合口过大常常引起肺水肿、肺动脉高压且行根治术时难以拆除吻合血管，现已较少应用[10]。1955 年 Davidson 完成了升主动脉－主肺动脉侧侧分流术，即 CS，CS 有效提升了肺血灌注，在促进肺血管发育，提升氧饱和度上作用明显，同时手术时操作简单，但其远期死亡率高、肺动脉扭曲发生率高、在后续行根治术时不易于拆除且有时会因为肺过度灌注引起充血性心力衰竭，在 PTFE 管道问世后，1975 年 Gazzaniga 等人以 PTFE 管道进行升主动脉-主肺动脉双侧的端侧吻合替代了之前的方式，改良后相较之前易于拆除、便于控制分流量不容易造成充血性心力衰竭[11] [12]。1962 年 Waterston 完成了升主动脉－右肺动脉侧侧吻合术，即 Waterston 手术，这种术式改善了上肢的血液循环，降低了血栓形成的风险，但易造成右侧肺动脉的扭曲且不易拆除，后续利用 PTFE 管道改良的 Waterston 手术明显改善了这些缺点，因此在临床上获得了广泛应用[13]。

2.2. 根治手术

TOF 一旦发现，只要满足一期根治的条件，均应行手术治疗。早期手术有利于保护由心室功能，促进肺动脉的发育和生长，减少慢性低氧血症对于心脏和神经系统的损害。一期矫治的基本条件为肺动脉发育能够承接全部的心输出量，肺动脉发育指标：McGoon 比值 > 1.2、Nakata 指数 > 150 mm^2/m^2 [14]。

1954 年 Lillehei 团队首次进行了在受控交叉循环下经右心室肺入路 TOF 矫治术[15]，106 名患者术后平均 23 年的随访中，存活率为 80% [16]。同年，Kirklin 首次应用人工心肺机进行四联症的根治手术也获得成功[17]，110 名患者术后 5 年随访生存率为 83%，同时多数患者存在的肺动脉瓣反流被认为意义不大[18]。一开始，人们认为 TOF 的修复需要完全剪除右室流出道增生的肌束，不留下任何残余梯度，因此采用右心室入路修补室间隔缺损，剪除增生的肌束以彻底疏通右心室流出道，如果肺动脉瓣超过轻度狭窄则将纵行切口跨过肺动脉瓣延长至主肺动脉，最后用心包补片修补切口。这样激进的策略在后续带来了一系列的问题，术后短期或长期的心律失常、右室功能障碍、右室扩张及严重的肺动脉瓣反流[19]。1981 年，人们意识到某些心脏传导束从那些增生的肌束中通过，切除这些肌束会影响心脏收缩功能及造成心律失常，因此建议保留这些肌束[20]。1962 年 Hudspach 首次报道经右心房径路做四联症的矫治手术，在 1985 年和 1987 年 Kawashima Y、Kavey RE 等人都进行了右心房联合肺动脉入路的尝试，此径路避免了右室切口，具有保护右心室功能和减少术后心律失常的优点[21] [22]。直到 1995 年，人们认识到保留肺动脉瓣的重要性，肺动脉瓣反流会带来右室扩张和右心室功能障碍[23]。因此，在进行 TOF 根治时，策略转变为尽量保存完整的肺动脉瓣，即使因为肺动脉瓣过于狭窄不得不做跨环切开，也尽量植入心包单瓣补片或 PTFE 单瓣补片以减少或延缓肺动脉瓣反流。多项研究证实，相较跨环补片，单尖瓣植入在减少术后肺动脉瓣反流上有显著的优势，但在长期的随访中人工材料补片或自体心包补片容易钙化且无法生长造成了早期或晚期植入瓣膜的功能障碍[24] [25] [26] [27] [28]。在疏通右室流出道时，策略转变为在室上脊附近尽可能少的剪除肌肉，并接受交界性肺动脉瓣狭窄，允许不超过 40 mmHg 的残余梯度存在，以避免持续的肺动脉瓣反流造成的容量负荷[29]。从上世纪 50 年代至今，随着体外循环手术修复技术的

标准化和进步，以及更好的术后管理，人们 TOF 的认识有所增加，围术期死亡率低至 3% [30]，存活率提高到了 85%~90% [31] [32]。

3. 并发症

3.1. 短期并发症

术后即刻常见的并发症是残留室间隔缺损，以及持续的右室流出道梗阻[33]。TOF 根治术后可能出现心律失常，并有室性心动过速、房颤和房内折返性心动过速的风险。心电图通常表现为右束支传导阻滞或左束支传导阻滞伴复合性心动过速。修复术后的患者可能会出现心脏性猝死。快速性心律失常和心脏性猝死的危险因素包括修复时年龄较大、男性、术后第三天之后的一过性完全性心脏传导阻滞以及 QRS 持续时间大于 180 毫秒[34]。目前临床多采用肺动脉球囊扩张术或肺动脉瓣置换术治疗法洛四联症术后残余右心室流出道梗阻，可以缓解肺动脉分支狭窄[35]。

3.2. 长期并发症

TOF 根治术的长期并发症包括肺功能不全引起的右室容量超负荷、流出道补片或切开引起的右室壁瘤、远端肺动脉阻塞、室壁肥厚、腔扩大、双室功能障碍、以及主动脉根部扩张和关闭不全[36]。患者死亡的三个主要原因是心律失常、心力衰竭和再手术并发症[37]。手术 30 年后猝死的风险增加到 6%至 9%；与此相关的一些因素包括 QRS 持续时间大于 180 毫秒，修复时年龄较大(大于 3 年)，明显的肺动脉瓣或三尖瓣反流，晕厥病史，多灶性室性早搏和室性心动过速[38]。再次手术最常见的指征是肺动脉瓣反流，而肺动脉瓣置换术的标准是根据 MRI 或 CT 扫描上的返流分数来衡量严重程度。这些研究的参数包括右室和左室收缩末期和舒张期末容量指数、射血分数和造成血液流出梗阻的室壁瘤[39]。患者可能会有运动不耐受、心力衰竭的体征和症状、晕厥和持续性室性心动过速。经导管经肺动脉瓣入路也可行肺动脉瓣置换术[40]。

4. 结语

TOF 是最常见的先天性心脏病之一，从上世纪中叶开展矫治手术至今，随着对其疾病发展的认识及相关手术水平的不断提高，相关诊断技术及术中辅助技术蓬勃发展，术后的死亡率和并发症发生率也较前明显下降。本文结合国内外近年来相关研究，对 TOF 外科治疗的发展历史及术后并发症的应对策略做了简单阐述，但目前国内外对上述许多问题仍无统一认识，还需继续努力进行相关研究，为 TOF 患者的治疗提供更多理论及临床依据。

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