

doi: 10.13241/j.cnki.pmb.2018.04.025

15 例儿童心动过速性心肌病的诊疗体会*

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摘要 目的:探讨儿童完全性心动过速性心肌病(pTIC)的临床特点、治疗及预后。**方法:**回顾分析 2009 年 1 月至 2016 年 10 月安徽省儿童医院心内科收治的 15 例完全性心动过速性心肌病患儿的临床表现、心功能、心电图、心脏彩超的特点,观察心律失常控制后的心室率、心脏左室内径大小及心功能恢复情况。**结果:**15 例 pTIC 患儿以室上性快速心律失常多见(14 例),10 例单纯药物治疗,3 例接受射频消融转为窦性心律,2 例失访。随访半年至 3 年与治疗前比较心室率明显下降[(116±27)次/分 vs. (189±28)次/分]、NT-proBNP 降低[(404±355)pg/mL vs. (6280±3155)pg/mL]、心脏左室舒张末内径变小[(3.12±0.48)cm vs. (3.69±0.70)cm]、左室射血分数升高[(57.9±9.3)% vs. (42.2±9.5)%]、改良 ROSS 评分下降[1(0-5)分 vs. 7(4-10)分]。**结论:**儿童 pTIC 由各种快速心律失常引起,心脏扩大和心功能障碍可完全恢复,早期识别、有效治疗的儿童 pTIC 长期预后良好。

关键词:快速性心律失常;心肌病;儿童

中图分类号:R542.2;R725.4 **文献标识码:**A **文章编号:**1673-6273(2018)04-718-03

Clinical Diagnosis and Treatment Experience of 15 Cases of Children with Tachycardia-Induced Cardiomyopathy *

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ABSTRACT Objective: To analyze the clinical features, efficiency of anti-arrhythmic medications and prognosis of children with tachycardia-induced cardiomyopathy (pTIC). **Methods:** The clinical data of 15 cases of children with pTIC admitted in the children's hospital of Anhui province from January 2009 to October 2016 were retrospectively analyzed and followed up. The clinical features, cardiac function, electrocardiographic and echocardiographic evaluation were analyzed. The ventricular rate, the size of the left ventricular diameter and the recovery of cardiac function were observed after cardiac arrhythmia was controlled. **Results:** The supraventricular arrhythmia was the commonest arrhythmia in 15 cases of pTIC (14 cases), there were 10 cases of pure drug therapy, 3 patients received radiofrequency ablation, and 2 cases were lost. The ventricular heart rates of was decreased compared with those of pretherapy [(116±27)bpm vs. (189±28)bpm], NT-proBNP was decreased[(404±355)pg/mL vs. (6280±3155)pg/mL], left ventricular end-diastolic diameter was shrunken [(3.12±0.48)cm vs. (3.69±0.70)cm], ejection fraction was increased[(57.9±9.3)% vs. (42.2±9.5)%], and Modified Ross score as decreased [1 vs. 7] after six months to 3 years. **Conclusions:** PTIC was caused by a variety of rapid arrhythmias. Heart enlargement and cardiac dysfunction could be fully restored. Children given early recognition and effective treatment had better prognosis.

Key words: Tachyarrhythmia; Cardiomyopathy; Child

Chinese Library Classification(CLC): R542.2; R725.4 **Document code:** A

Article ID: 1673-6273(2018)04-718-03

前言

心动过速性心肌病 (Tachycardia-Induced Cardiomyopathy, TIC) 是持续或反复发作的快速心律失常导致的以严重的心脏扩大、心室壁变薄和心室收缩功能减低为主要表现的可逆性心脏病^[1]。Fenelon 等根据患者是否合并器质性心脏病将 TCM 分为非单纯型 TIC 与单纯型 TIC(pure TIC, pTIC)。pTIC 类似于扩张型心肌病,不同的是当快速性心律失常和心力衰竭得到控制后,心脏可以完全恢复正常^[2]。

自 1913 年 Gossage^[3]首次提出心房颤动并发心功能下降及心脏扩大患者类似扩张型心肌病样的临床表现,相继出现不少 TIC 的报道,但大多为成年慢性房颤患者,有关儿童 TIC 的报道较为零散。儿童长期反复发作的快速心律失常比成人更易出现心力衰竭及心脏扩大,治疗不及时可能发展成扩张性心肌病。有学者报道^[4]即使当心功能恢复后仍然有患者发生猝死,心律失常复发后左室功能迅速恶化,所以需对 pTIC 高度警惕,对患者的密切随访颇为重要,但目前暂无 TIC 的诊疗指南。为增强对本病的认识,避免漏诊误诊,提高诊治水平,现将本院

* 基金项目:安徽省卫生厅科研基金项目(13FR020)

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(收稿日期:2017-08-16 接受日期:2017-09-11)

2009年1月至2016年10月收治的15例pTIC患儿的临床资料总结报道如下。

1 资料和方法

1.1 一般资料

选择我院2009年1月至2016年10月收治的pTIC患儿15例,男13例,女2例,发病年龄1~12月13例,平均4个月;2岁1例;13岁1例。纳入标准为:快速性心律失常,平均心室率超出同年龄的正常值;临床表现为心功能不全,伴有心脏扩大,超声心动图左室舒张末期内径(left ventricular end-diastolic diameter, LVDD)超出正常值,并且左室射血分数(ejection fraction, EF) <55%;控制心室率后,心功能及心脏大小恢复或明显好转,EF>55%。排除心肌炎、扩张型心肌病(心律失常纠正后LVDD、EF无明显改善)及结构性心脏病。

1.2 治疗

本组的主要治疗包括抗心律失常药物(常用Ic类普罗帕酮、II类美托洛尔、III类胺碘酮、索他洛尔)和控制心衰药物(洋地黄类、利尿剂、血管紧张素转换酶抑制剂)治疗。药物抗心律失常不理想时选用射频消融术。根据每个患儿情况选择抗心律失常药及改善心功能药物,首选普罗帕酮静推,心率控制不理想予胺碘酮5 μg/Kg维持,心室率控制仍不理想时加用美托洛尔或改用索他洛尔^[9],出现心功能不全患儿先予西地兰饱和,然后地高辛维持。阵发性室上性心动过速患儿予ATP终止发作,伴肝大患儿予呋塞米、氢氯噻嗪利尿。以24小时动态心电图平均心率为评判标准,控制心室率在正常范围。

1.3 随访

控制心动过速和心力衰竭,患儿病情稳定后出院,定期于我科门诊随访。

1.4 统计学分析

应用SPSS17.0统计软件处理数据,计量资料符合正态分布的用平均数±标准差表示,不符合正态分布的用中位数表

示,分别采用配对t检验和配对秩和检验对数据进行分析,以P<0.05为差异有统计学意义。

2 结果

2.1 临床表现

起病隐匿,临床症状轻重不一。病史2天至1个月不等,从发现心律失常到心功能不全的临床表现平均时间7天,入院时平均心率189次/分,舒张末期左室内径3.68 cm,左室射血分数42.2%,血清氨基末端B型钠尿肽原(NT-proBNP)6280 pg/mL,改良ROSS评分中位数7分。

15例患儿中,13例为小于1岁婴儿,1例2岁幼儿,1例13岁青春期儿童。入院时临床表现:11例为发现心动过速,9例肝脏肿大,9例全胸片显示心影增大,7例表现为气促,7例食纳差,1例发现水肿伴少尿。超声心动图15例均显示心脏不同程度扩大和左心收缩功能减低,7例二、三尖瓣关闭不全伴轻度反流。心电图示7例房性心动过速,3例房扑,2例阵发性室上性心动过速合并预激,1例结性心动过速,1例交界性心动过速,1例室性早搏和短阵室速,均无器质性心脏病。

2.2 治疗效果

所有患儿均首选药物治疗,心室率均较入院前明显下降,其中10例单纯药物治疗完全转为窦性心律;5例患儿快速心律失常频繁发作,其中3例于外院行射频消融术后转为窦性心律,2例转院后失访。9例心脏大小恢复正常,6例患儿左心室仍稍扩大,有12例患儿心功能恢复正常。

治疗前平均心率189±28次/分,超声心动图LVDD平均3.69±0.70 cm,EF值42.2±9.5%,NT-proBNP 6280±3155 pg/mL,改良ROSS评分中位数7(4-10)分。治疗后平均心率116±27次/分,超声心动图LVDD平均3.12±0.48 cm,EF值57.9±9.3%,NT-proBNP 404±355 pg/mL,治疗前后差异有明显统计学意义(P<0.05),见表1;改良ROSS评分中位数1(0-5)分,差异有明显统计学意义(P<0.05)。

表1 治疗前后各指标比较($\bar{x} \pm s$)

Table 1 Comparison of indexes pre-therapy and post-therapy($\bar{x} \pm s$)

Groups	Heart Rate	NT-proBNP (pg/mL)	LVDD(cm)	EF(%)
Pre-therapy	189±28	6280±3155	3.69±0.70	42.2±9.5
Post-therapy	116±27*	404±355*	3.12±0.48*	57.9±9.3*

Note: *P<0.05 vs. Pre-therapy.

2.3 随访情况

13例患儿获得随访,失访2例。13例随访患儿中,5例心动过速仅发作1次,另8例患儿现一般情况良好,其中2例婴儿和1例13岁患儿积极抗心律失常后心室率虽有所减慢,但时有室上性心动过速发作,建议其转院行射频消融术,经射频消融术后患儿转为窦性心律;但其中1例仍时有心律失常发作,现已随访2年,继续口服索托洛尔,心功能恢复正常。

3 讨论

pTIC是一种罕见的可逆性左心室收缩功能障碍的病因,易误诊为扩张型心肌病和心力衰竭^[6,7]。快速心律失常包括心房

颤动、心房扑动、房性心动过速、房室折返性心动过速、房室折返性心动过速及室性心动过速等^[8,9]。持续性交界区折返性心动过速常见于婴幼儿,心房颤动、心房扑动或持续性室性心动过速尽管多见于老年患者,但其也能导致儿童pTIC^[10]。其诊断较困难,依赖于心率控制之后临床症状、心功能及左室内径的改善^[11]。因此,同时存在快速心律失常、心功能减低及心脏扩大的患儿不必急于诊断,积极抗心律失常治疗及控制心室率后再下定论。

本研究纳入的15例患儿中,14例为室上性心动过速,仅1例室性心动过速,其中10例为房性心动过速,与Houmsse M^[12]等认为慢性房性心动过速是儿童TIC的最常见病因符合。pTIC

的典型临床表现类似于其他原因引起的心力衰竭,年龄越小的患儿,临床表现越缺乏特异性^[13]。本组 15 例患儿中,13 例为小于 1 岁的婴儿,入院时临床表现为呼吸急促、多汗、喂养困难,年长儿表现为气促、运动不耐受、劳力性呼吸困难、胸痛、心悸、晕厥,与 Ramesh^[14]和 Macicek 等^[15]的报道相同。心室功能障碍程度常常与心动过速的持续时间和速度有关^[16],如果心率增快的较慢,心动过速开始后几周至几个月年长儿童和青少年才表现出心力衰竭^[17],而婴儿通常在几天后就出现心衰的症状并且无法用言语表达不适,猝死的风险可能会高达 10%^[18],需高度警惕。因此,尽早发现潜在的快速性心律失常可减少 pTIC 的发生及猝死的可能性。

pTIC 的诊断较困难,目前尚无专家共识及诊疗指南,依赖于心率控制之后临床症状的改善,发生心律失常和心力衰竭的先后顺序^[19,20]。对于已经出现心力衰竭的患儿,需要尽早发现潜在的快速性心律失常。本组 11 例患儿因发现心率快入院,心电图不仅可提示心律失常类型,还可提示心脏扩大,或伴有窦性心动过速,非特异性 ST-T 波变化,心室肥厚等表现^[21]。超声心动图心室收缩功能障碍通常是 pTIC 患儿首先表现,紧随其后的是左心室扩张^[22],并且很可能出现左室进行性扩大,所以动态监测心脏彩超必不可少。

控制快速心律失常是 pTIC 转归的关键^[23]。本组 15 例患儿治疗前左室内径均较同年龄儿童大,左室收缩功能减低,治疗后心动过速改善显著,左室收缩功能基本恢复正常,但只有 9 例患儿左室内径正常,6 例患儿左室内径和左室容积仍然较正常稍大,这与韩晓华等的研究相同^[24]。心功能的恢复同样取决于心律失常的心室率和持续时间两个因素,婴幼儿大多可完全恢复。心功能最早在一周内可恢复,完全恢复约 4-6 周。BNP 是反映心功能的敏感指标^[25],所有患儿 BNP 均升高,积极抗心律失常及抗心衰治疗后 2 周,BNP 明显下降,随访 1 月后基本恢复正常。但心功能恢复正常的 pTIC 患儿与同年龄正常儿童心脏彩超参考值对比,pTIC 患儿左室内径大,进一步说明即使心功能恢复正常,LVDD 恢复正常大小的时间仍需长期随访。

目前,国内外对儿童 pTIC 的相关报道不多,齐建光^[26]等报道儿童 pTIC 药物治疗有效率为 (8/12),4/12 患儿需射频消融。Hayrettin 等^[27]报道 12 例 TIC 患儿,药物治疗后 4 例复发,3 例射频消融成功,1 例失败需安装永久起搏器。本组 15 例患儿均首选药物治疗,所有患儿心率均较入院前明显下降,其中 10 例单纯药物治疗完全转为窦性心律,与之相似。3 例患儿房性心动过速频繁发作,于外院射频消融术后转为窦性心律。余 2 例药物治疗效果不佳,建议转院行射频消融术后失访。获得随访的患儿中,9 例心脏大小恢复正常,均为小于一岁的婴儿,与 Jeremy 等^[28]认为年龄越小的儿童心脏恢复正常的可能性越大相同。

总之,当患儿出现快速性心律失常,特别是房性心律失常,同时伴有心脏扩大,心功能不全时,需考虑本病,并尽早纠正心律失常。如能做到早期诊断及治疗,将大大改善其预后。治疗有效的儿童 pTIC 通常长期预后良好,而对药物不敏感或射频消融术后仍有反复快速心律失常的 pTIC 更需密切随访。

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