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·病例报道·

Echocardiographic diagnosis of Raghib syndrome: a case report

超声心动图诊断Raghib综合征1例

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[中图法分类号]R540.45;R825.4 [文献标识码]B

患儿男,1岁,因发现心脏杂音1年就诊。体格检查:心界向左扩大,心率76次/min,律齐;胸骨左缘第二三肋间可闻及Ⅲ期/6级收缩期杂音,肺动脉瓣区第二心音亢进,呈固定分裂。心电图示:窦性心律。胸部X线检查:心影增大。超声心动图检查:右心增大(右室内径15 mm,右房内径24 mm),肺动脉增宽(内径15 mm);非标准主动脉短轴切面显示一异常血管汇入左房近左心耳处,向上追踪扫查确定该血管为永存左上腔静脉(PLSCV),右上腔静脉汇入右房。冠状静脉窦及无名静脉缺如。房间隔中份回声失落约16 mm,心房水平左向右分流;大血管水平左向右为主双向分流,分流束宽1.5 mm。见图1 A~E。超声心动图提示:先天性心脏病房间隔缺损(继发孔型),双上腔静脉,PLSCV汇入左房,动脉导管未闭(细小管型),肺动脉高压。增强CT及三维重建检查:粗大左颈内静脉通过左上腔静脉汇入左房;右侧上腔静脉汇入右房;左、右上腔静脉间未见粗大交通支。见图1F。提示:双上腔静脉,左上腔静脉直接汇入左房。患儿于全麻体外循环下行永存左上腔静脉异位引流矫治、房间隔缺损修补及动脉导管结扎术,术中所见证实超声心动图提示。术后患儿恢复良好。

讨论:Raghib综合征是一种罕见的先天性心脏病,系指PLSCV直接开口于左房,同时合并冠状静脉窦缺如和房间隔缺损;发病率不到房间隔缺损总数的1%[1],临床较为罕见。Raghib综合征是胚胎发育的过程中左房静脉皱褶和左主静脉发育不良,造成冠状静脉窦与左房之间的共同壁完全缺如,属于无顶冠状静脉窦综合征中的一类[2]。超声心动图是最常用于诊断Raghib综合征的无创检查方法,其表现易与单纯房间隔缺损混淆,因此常规观察冠状静脉窦对于提示PLSVC的存在有重要意义。但因PLSVC直接汇入左房者较罕见,检查过程中容易漏诊,亦误诊为左上肺静脉,检查时需结合多切面进行观察,特别是胸骨上窝切面。本例患儿超声诊断要点是在左房近左心耳处侧壁可见一异常粗大静脉汇入,再追踪扫查发现其为PLSVC直接汇入左房,同时存在房间隔缺损和冠状静脉窦缺如。对于超声图像质量不佳者,可通过增强CT检查或心血管造影明确诊断。

Raghib综合征需要进行手术矫治,常规行外科开胸手术,将PLSVC吻合到右房;亦有文献[3]报道应用心血管造影经皮治

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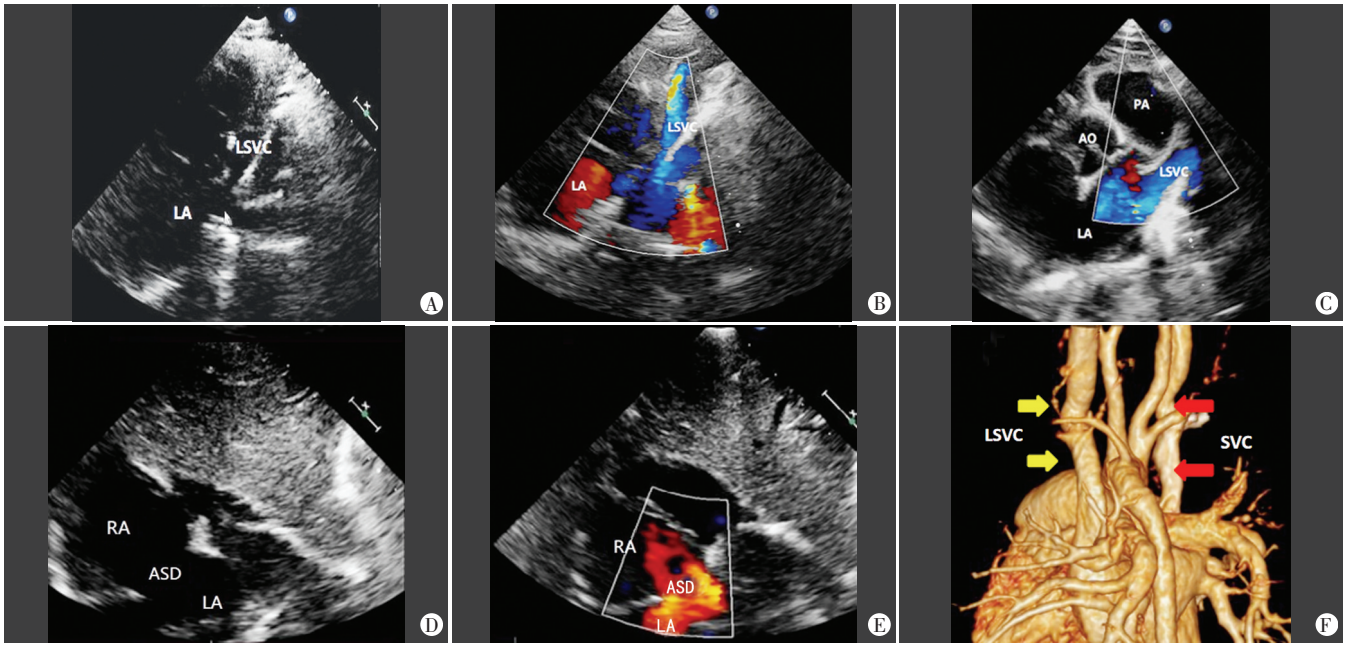

A:二维超声显示左上腔静脉直接开口于左房;B、C:彩色多普勒显示左上腔静脉向下走行的血流,最终汇入左房;D、E:剑突下双房心切面显示房间隔缺损及房水平分流;F:CT三维重建示永存左上腔静脉直接汇入左房(黄色箭头示),右上腔静脉汇入右房(红色箭头示),左、右上腔静脉间未见交通支。LSVC:左上腔静脉;LA:左房;AO:主动脉;PA:肺动脉;RA:右房;ASD:房间隔缺损;SVC:右上腔静脉

图1 Raghib综合征超声图像及CT三维重建图

疗。本例患儿因合并较大的房间隔缺损,以及动脉导管未闭,因此行外科矫治术。术后患者恢复良好。

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