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# 几种少见的风湿性疾病的心血管表现

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**【摘要】** 一些常见的风湿免疫性疾病,如类风湿关节炎、干燥综合征以及特发性炎症性肌病常合并心血管损伤,这些心血管病变甚至是风湿病最常见的死亡因素。其他的一些不常见的风湿免疫性疾病,如成人 Still 病、未分化结缔组织疾病以及结节性脂膜炎等是否也合并了一些常见的心血管疾病,目前不得而知。现探讨以上几种少见的风湿病的心血管表现。

**【关键词】** 成人 Still 病; 未分化结缔组织病; 结节性脂膜炎; 心血管疾病
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# Cardiovascular Manifestations in Rare Rheumatic Diseases

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**【Abstract】** Some common rheumatic diseases, such as rheumatoid arthritis, Sjogren's syndrome, and idiopathic inflammatory myopathies, often coexist with cardiovascular diseases, which also are the most common factors of death. However, whether other rheumatic diseases including adult-onset Still's disease, undifferentiated connective tissue disease and panniculitis, have the same features or not, is still unknown. Our paper aims to review the cardiovascular manifestations of these rare rheumatic diseases.

**【Key words】** Adult-onset Still's disease; Undifferentiated connective tissue disease; Nodular panniculitis; Cardiovascular disease

常见的一些风湿免疫性疾病,如类风湿关节炎、干燥综合征以及特发性炎症性肌病常合并循环系统

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的损伤[1-3]，但其他的一些风湿免疫性疾病，如成人 Still 病（adult-onset Still's disease，AOSD）、未分化结缔组织疾病（undifferentiated connective tissue disease，UCTD）、结节性脂膜炎（nodular panniculitis，NP）等，如按照风湿免疫性疾病的炎症特点，可能并存心血管疾病及相关危险因子。然而相关研究很少，关于以上疾病的心脏受累症状及表现，中华医学会相关指南及国外指南均未提及。那么，诸如心肌炎、心力衰竭、冠心病以及代谢紊乱是否与以上疾病共存呢？

## 1　AOSD

Still 病本是指系统型起病的幼年型慢性关节炎，相似的疾病发生在成人故称之为 AOSD。既往有认为本病是一种介入风湿热与幼年型类风湿关节炎之间的变应性疾病，是类风湿关节炎的一个疾病过程[4]。AOSD 男女患病率相似，散布世界各地，好发年龄为 16～35 岁。其病因及发病机制尚不明确。免疫紊乱、感染、遗传、精神因素、变态反应均与之相关。临床特点表现为发热、关节痛和/或关节炎、皮疹、中性粒细胞增多，严重者可伴系统损害。

AOSD 诊断无特异方法，主要基于排他性诊断的基础上，因为其他风湿免疫性疾病如系统性红斑狼疮、类风湿关节炎、强直性脊柱炎等早期阶段，可出现酷似 AOSD 的表现；另外，一些肿瘤的早期阶段也可出现 AOSD 的表现。非甾体类抗炎药物、糖皮质激素以及抗风湿药物仍是治疗 AOSD 的标准用药，少数患者呈自愈倾向，多数患者反复发作，迁延不愈。

据文献报道[5-8]，心包积液是 AOSD 较为常见的心血管合并症，约 1/4 的 AOSD 患者合并心包积液，少量的心包积液可自主吸收，稍多的心包积液通过糖皮质激素、抗风湿药物以及丙种球蛋白的作用也可吸收。较大量的心包积液常常和心包压塞相关，迄今为止，有 15 例 AOSD 合并心包压塞的病案报道。心包压塞作为一种危及患者性命的病症，临床需要注意其症状，如呼吸困难、胸痛、咳嗽等，心脏超声检查及在其引导下细针穿刺是必须的。

心肌炎的病案报道较多，涉及面很广，根据目前的可利用资料[9-14]，心肌炎可能也是 AOSD 的一类重要的心肌损害表现，AOSD 合并心肌炎可能并不少见，有时会成为 AOSD 的首发表现，即使临床治愈，也可能复发，同样，心肌炎也可能成为 AOSD 的致死性原因。在部分病例中，糖皮质激素或者抗风湿药物对心肌炎有较好的疗效，如果这两类药物疗效不佳，也可以考虑丙种球蛋白、阿那白滞素以及肿瘤坏死因子拮抗剂等治疗，部分患者也可获得满意的疗效。尽管这些生物制剂可能有致心力衰竭的潜在风险，但这一点尚未定论，根据生物制剂在类风湿关节炎及其他风湿结缔组织疾病中的经验，总体来看，生物制剂减少风湿病的总体死亡率及心血管病死率。Yang 等[15] 在 2008 年报道了 1 例 AOSD 合并充血性心力衰竭的患者经肿瘤坏死因子拮抗剂治疗后，心力衰竭症状及左室射血分数好转的病例。

AOSD 常会导致心肌炎，按常理推论，可能会使心功能受损，严重时可导致心力衰竭。部分作者报道了 AOSD 合并心功能受损的病案[9-10,16]，Ueda 等[17] 解释心功能受损可能与心肌微血管的病变有关。此外，AOSD 合并二尖瓣及主动脉根部病变及肺动脉高压也有报道，这类患者的预后较差[18-20]。

## 2　UCTD

UCTD 是一类具有结缔组织疾病的常见表现，如关节炎、雷诺现象、肌炎、浆膜炎、肺间质病变等，但又不符合目前已知的任何一种特定的结缔组织疾病的诊断标准。LeRoy 于 1980 年提出，直到 1989 年相关临床研究才逐渐增多，目前认为 UCTD 可能属于某一种弥漫性结缔组织疾病的早期阶段或顿挫型，部分患者也可能是一种独立的阶段[21-22]。

关于 UCTD 的心血管病变报道非常少。目前，有 2 例病案报道涉及了 UTCD 的心包压塞，心包穿刺后，经常规抗风湿药物治疗后患者症状明显好转[23-24]。此外，Louthrenoo 等[25] 报道了 1 例抗 Ro/SSA 抗体阳性的孕妇生育了 1 例先天性的完全性房室传导阻滞的婴儿，这提示抗 Ro/SSA 抗体对 UCTD 的心脏传导系统可能有一定影响。实际上，UCTD 患者的先天性传导阻滞远较系统性红斑狼疮常见，可能与抗 Ro/La 抗体阳性相关[26]。

最近，Laczik 等研究者发现 UCTD 的患者血管内皮细胞存在持续的损伤，氧化的低密度脂蛋白以及炎症反应可能参与这个过程，并与颈动脉内膜增厚相关，这可能提示与其他风湿病一样[27]，动脉粥样硬化也参与到 UCTD 患者的心血管损伤中[28]。

## 3　NP

NP 是一种原发于脂肪小叶的非化脓性炎症。又称之为韦伯病（Weber-Christian disease）。病因不明，可能与脂肪组织异常代谢及免疫损伤有关。本病多发于女性，约占 75%，好发于 30～50 岁人群。临床起病呈急性或亚急性过程，以反复全身不适、关节痛、发热、皮下结节为特征。

根据中华医学会风湿病学分会诊治指南意见，NP 的诊断主要依据以下几点：好发于青壮年女性；以反复发作与成批出现的皮下结节为特征，结节有疼痛感和显著触痛，消退后局部皮肤出现程度不等的凹陷和色素沉着；常伴发热、关节痛与肌痛等全身症状；当病变侵犯内脏脂肪组织，视受累部位不同而出现不同症状。内脏受累广泛者可出现多脏器功能衰竭、大出血或并发感染。糖皮质激素加免疫抑制剂的治疗仍然是 NP 的标准治疗方法，当病变累及内脏时预后较差[29]。

心力衰竭可能是 NP 的一个典型的心血管合并症，超声心动图异常可表现为左心增大、左室壁运动弥漫性减弱、左室射血分数减低；尸检发现 NP 患者心肌细胞变性、心肌间质纤维化；除累及心肌外，也可累及心包，但具体机制尚不明确[30-32]。

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