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·综述·

儿童寻常型天疱疮的疾病特征和管理

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【摘要】 寻常型天疱疮是最常见的天疱疮亚型，好发于成人，儿童患者较为罕见。尽管由于临床治疗手段的发展，儿童寻常型天疱疮的死亡率已呈现逐步下降的趋势，但在缺乏正确诊断以及治疗手段的情况下，其死亡率仍可高达70%～100%。对于发病较急且病情进展迅速的患儿，及时获得适当的治疗能避免出现严重后果。然而，由于疾病的罕见性，目前缺乏儿童寻常型天疱疮的诊断和治疗指南。本文对1969至2024年间Pubmed及中国知网数据库收录的104例儿童寻常型天疱疮患者进行综述，以期为儿童寻常型天疱疮的规范化诊疗提供参考。儿童寻常型天疱疮较为罕见，病损表现为皮肤、黏膜松弛性水疱，疱破后遗留鲜红色糜烂面。文献中报道的病例约占所有寻常型天疱疮病例的1.4%～3.7%，其发病年龄为1.5～18岁，平均12.4岁，男女无明显差异。儿童患者的口腔黏膜通常最早受累，累及率高达87.3%，其中颊部（27.9%）累及率最高；其他黏膜累及率达52.9%，其中生殖器（28.8%）和肛周（6.7%）累及率高于成人患者。皮肤损害累及率达80.4%，高于成人患者（16.0%～68.4%）。若病损较为局限，可先尝试局部糖皮质激素治疗，8.3%的患儿仅采用局部治疗可达完全缓解。若对局部治疗反应不佳，首选糖皮质激素系统治疗，75.3%的患儿使用了泼尼松，85.1%的泼尼松起始口服剂量为0.5～1.5 mg/kg/d，14.9%采用了2 mg/kg/d的初始剂量。对糖皮质激素治疗反应不佳或不良反应严重的患儿，可考虑使用免疫抑制剂、生物制剂以及其他药物辅助治疗，使用率较高的药物主要是硫唑嘌呤（24.0%）、氨苯砜（21.7%）和利妥昔单抗（12.5%）。患儿随访时间为1～120个月，平均38个月，其预后优于成人患者，43.8%的患儿达到完全缓解（停止治疗），37.5%达到部分缓解（低剂量维持），9.6%仍在接受治疗，仅有1.1%死于肺炎和败血症。相较于成人，儿童长期服用糖皮质激素，可能会对生理功能和精神心理造成不良影响，更容易出现生长发育、代谢及眼部不良反应。22.1%的患儿在使用激素后出现了严重不良反应，其中库欣样面容（73.9%）和体重增加（39.1%）最为常见，30.4%的患儿出现了生长与骨骼发育的不良反应，包括生长迟缓（17.4%）、骨质疏松（8.7%）和骨折（4.3%）。儿童寻常型天疱疮与成人患者的病因、临床特点和组织病理学特点具有一定的相似性，关键在于早期诊断，及时治疗，必要时通过多学科团队合作及时进行干预，保护患儿身心健康。

【关键词】 儿童寻常型天疱疮；　儿童自身免疫性大疱病；　桥粒芯糖蛋白；　直接免疫荧光；　间接免疫荧光；　糖皮质激素；　利妥昔单抗；　硫唑嘌呤

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Characteristics and management of pediatric pemphigus vulgaris

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【**Abstract**】 Pemphigus vulgaris (PV) is the most common subtype of pemphigus. It predominantly affects adults, with pediatric cases being exceedingly rare. Despite advancements in clinical treatment, the mortality rate of pediatric PV (PPV) has historically been alarmingly high, ranging from 70% to 100% in the absence of proper diagnosis and treatment. Although recent improvements in therapeutic strategies have led to a gradual decline in mortality, early and appropriate intervention remains crucial, particularly for children with acute onset and rapid disease progression, to prevent severe complications. However, due to the rarity of PPV, no standardized diagnostic and treatment guidelines are currently available. This study retrospectively analyzed 104 PPV cases recorded in the PubMed and China National Knowledge Infrastructure (CNKI) databases between 1969 and 2024, with the aim of providing insights for the standardized diagnosis and management of PPV. PPV presents with flaccid blisters affecting both cutaneous and mucosal surfaces. Upon rupture, these blisters result in painful, sharply demarcated erythematous erosions, accounting for approximately 1.4% - 3.7% of all reported PV cases. The age of onset ranges from 1.5 to 18 years, with an average of 12.4 years, and no significant gender differences have been observed. In pediatric patients, the oral mucosa is typically the earliest and most frequently affected site, with an involvement rate as high as 87.3%, and it most commonly affects the buccal mucosa (27.9%). Other mucosal sites are affected in 52.9% of cases, with genital (28.8%) and perianal (6.7%) involvement being more frequent than in adult patients. Skin lesions are present in 80.4% of pediatric cases, a significantly higher rate than 16.0% - 68.4% observed in adults. If lesions are relatively localized, local glucocorticoid therapy can be attempted first, with 8.3% of children achieving complete remission through local treatment alone. Systemic glucocorticoid therapy is the preferred option for cases that respond poorly to local therapy. Among these cases, 75.3% of pediatric patients were treated with prednisone, with 85.1% starting at an oral dose of 0.5 - 1.5 mg/kg/day, while 14.9% received an initial dose of 2 mg/kg/day. Alternative treatments, such as immunosuppressants, biologics, or other adjuvant medications, may be considered for pediatric patients who exhibit an inadequate response to glucocorticoid therapy or experience severe adverse effects. The most commonly used agents include azathioprine (24.0%), dapsone (21.7%), and rituximab (12.5%). The follow-up period for pediatric patients ranged from 1 to 120 months, with an average duration of 38 months. Prognosis in pediatric patients was more favorable compared to adults, with 43.8% achieving complete remission (cessation of treatment), 37.5% achieving partial remission (low-dose maintenance therapy), 9.6% still undergoing treatment, and only 1.1% succumbing to pneumonia or sepsis. Compared to adults, prolonged corticosteroid use in children poses a greater risk to physiological and psychological well-being, making them more susceptible to adverse effects related to growth, metabolism, and ocular health. Severe adverse reactions occurred in 22.1% of pediatric patients receiving corticosteroids, with Cushingoid facies (73.9%) and weight gain (39.1%) being the most common. In addition, 30.4% experienced growth and skeletal abnormalities, including growth retardation (17.4%), osteoporosis (8.7%), and fractures (4.3%). While PPV shares certain etiological, clinical, and histopathological characteristics with adult PV (APV), early diagnosis and timely intervention remain critical for optimal outcomes. Multidisciplinary collaboration is often necessary to ensure comprehensive management, improve treatment adherence, and safeguard the physical and psychological health of pediatric patients.

【**Key words**】 pediatric pemphigus vulgaris; pediatric autoimmune bullous diseases; desmoglein; direct immunofluorescence; indirect immunofluorescence; glucocorticoid; rituximab; azathioprine

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天疱疮是一种自身免疫大疱性疾病，其特点是产生针对桥粒芯糖蛋白（desmoglein，Dsg）的自身抗体，从而导致棘层松解，产生上皮内疱[1-2]。寻常型天疱疮（pemphigus vulgaris，PV）是其中最常见的亚型，约占所有病例的70%[3]。PV好发于成人，儿童患者较为罕见[4]。儿童PV（pediatric PV，PPV）是指发生于18岁及以下的PV，根据发生的年龄，可进一步划分为出生时或出生2周内发病的新生儿型天疱疮（newborn PV，NPV），12岁以上的青少年型天疱疮（juvenile PV，JPV），12岁以下的童年型天疱疮（childhood PV，CPV）[5]。

与成人PV（adult PV，APV）类似，PPV病损通常会累及口腔黏膜[6]。在缺乏正确诊断和治疗手段的情况下，其死亡率高达70%～100%[7]。由于

PPV的罕见性以及口腔黏膜病损的高发性，口腔医生的正确诊断能避免延误治疗时机。目前尚无PPV管理指南，本文将对PPV的流行病学、病因、临床表现、诊断、治疗、预后特点等方面进行综述，以期为PPV的规范化诊疗提供参考。

1　流行病学特点

PV的全球发病率为每年0.098/10万～5/10万[8-11]。PPV较为罕见，难以获得准确的发病率和患病率。文献中报道的PPV约占所有PV病例的1.4%～3.7%，其中CPV型较JPV更罕见[12-13]。

笔者总结了1969至2024年间Pubmed及中国知网数据库收录的关于PPV的中英文文献，共76篇113例患者，其中包括9例NPV。NPV通常是由于母体患有PV，抗体经胎盘转移导致新生儿罹患PV[14]。NPV具有自限性，出生后数周内可随着来自母体的自身抗体的清除而缓解，无需系统治疗[15-17]。除去NPV，PPV的发病年龄为1.5～18岁，平均发病年龄为12.4岁，男女比例为53∶51；发病高峰期为12～18岁，共73例（70.2%），12岁以下患者共31例（29.8%）。

2　病　因

儿童PV的病因与成人PV相似，尚未完全阐明，可能与以下因素有关。

2.1　遗传因素

PV具有一定的遗传基础，呈现地域和种族分布特点[18]，例如在德裔犹太人和地中海地区血统的人群中，PV的发病率较高。这一现象与多个人类白细胞抗原（human leukocyte antigen，HLA）II类基因密切相关，特别是HLA-DRB1*04和HLA-A*10[11,19]。此外，ST18（suppression of tumorigenicity 18）基因编码的促凋亡分子也与PV显著相关，尤其是在犹太人和埃及人群中[20]。

2.2　环境因素

遗传因素可能决定个体对PV的易感性，当易感个体暴露于已知的（诱发性PV）或未知的（特发性PV）诱发因子时，可能会导致PV爆发。许多外源性因素，如药物（青霉胺、青霉素、卡托普利、非甾体类抗炎药等）[21]、疫苗接种[22-23]、物理因素（烧伤、电离辐射）、病毒[24]或食物都可能诱发PV[25]。大部分病因尚不明确，在笔者总结的104例PPV患儿中，仅有4例（3.8%）可能为诱发性，其中3例与病毒感染有关[7, 26-27]，1例与使用药物有关[28]，其余病例的病因尚不明确。

3　临床特点

3.1　病损特点

PPV的病损表现为皮肤、黏膜松弛性水疱，疱破后遗留鲜红色糜烂面[29]。APV[30-35]与PPV的病损特点如下：PPV的口腔黏膜通常最早受累，累及率高达87.3%，其中颊部（27.9%）、唇部（25.0%）、牙龈（19.2%）、舌部（19.2%）发生率相对较高，此外还包括腭部（15.4%）和口底（2.9%）；其他黏膜累及率达52.9%，其中生殖器（28.8%）和肛周（6.7%）累及率高于APV，而咽喉（16.3%）和鼻腔（5.8%）累及率低于APV，其余可受累黏膜包括结膜（9.6%）和食管（3.8%）；皮肤损害通常在黏膜受累几周或几月后发生，累及率达80.4%，明显高于APV（16%～68.4%），好发于皮脂溢出区如面部（19.2%）、头皮（17.3%）、躯干（51.0%）和机械应力区如四肢（39.4%）。在102例明确发病部位的PPV患儿中，20例（19.6%）仅有黏膜病损，9例（8.8%）仅有皮肤病损，73例（71.6%）同时存在皮肤和黏膜病损，与APV无明显差异。

3.2　组织病理学和免疫学病理学特点

在笔者总结的104例PPV患儿中，有98例患儿进行了组织病理学检查，96例（98.0%）可见棘层松解、上皮内疱形成。病损局部的细胞涂片可用于协助诊断，其中可见存在特异性的Tzanck细胞，仅有15例患儿进行了细胞涂片检查，阳性率100%。

PPV诊断的免疫病理学试验可参考APV[36]，直接免疫荧光（direct immunofluorescence，DIF）是诊断PV最敏感和可靠的方法之一[37]，其阳性预测值接近100%[38]。文献报道的DIF阳性率为60%～100%[39-41]。在笔者总结的104例PPV患儿中，有93例患儿进行了DIF检查，其中92例（98.9%）显示棘细胞间有IgG和/或C3网状沉积。

间接免疫荧光（indirect immunofluorescence，IIF）检查可显示血清中的抗Dsg抗体在底物上皮细胞间网状沉积[42-43]，但其滴度并不总是与疾病严重程度相关，不能作为治疗的指导或监测疾病活动性的指标[44]。既往研究表明，70%～90%的PV患者的IIF呈阳性[45]。PPV患儿IIF阳性率稍高于APV患者，有58例患儿进行了IIF检查，其中53例（91.4%）阳性。

酶联免疫吸附测定法（enzyme-linked immunosorbent assay，ELISA）是诊断PV和病程免疫监测的

最佳血清学工具[46]，抗Dsg1和抗Dsg3自身抗体数值与PV的严重程度相关[47]。在新诊断的PV患者中，虽然Dsg3抗体存在于90%的患者中，但Dsg1抗体水平高于参考值的几率仅为46%～75%（≥20.0 U/mL）[48]。笔者总结的23例进行了ELISA检查的患儿中，全部表现为Dsg1和/或Dsg3高于参考值，19例明确了抗体类型，Dsg1抗体高于参考值的几率为73.7%，Dsg3抗体高于参考值的几率为100%。

在特殊情况下无法确定诊断时，需要更复杂的实验室检查（如免疫印迹，免疫沉淀）[49]。此外，HLA分型有助于在患儿基因型中寻找PV易感基因[20]。

4　诊断特点

PPV的诊断流程可参考APV，对于儿童出现口腔黏膜诱因不明的水疱、糜烂/溃疡，排除化学灼伤性口炎、药敏性口炎、急性疱疹性龈口炎等儿童常见口腔黏膜疾病后，应考虑大疱性疾病。组织活检是诊断大疱性疾病的必要检查，PPV患儿血清大疱性疾病特异性抗体的阳性率较高，对于不能配合的低龄儿童，可先行大疱性疾病特异性抗体筛查，必要时可在镇静或全身麻醉下进行组织活检。

5　治疗特点

5.1　治疗方案

PPV的治疗指南可参考APV患者。治疗目标是控制病损，同时尽可能减少治疗的不良反应。制定治疗方案时应综合考虑患儿年龄、体重、疾病严重程度和药物潜在不良反应等因素。

病损较为局限的轻度PPV，可先尝试局部涂擦或局部封闭糖皮质激素（glucocorticoid，GC）治疗。若局部治疗有效，通常持续应用至活动性皮损基本愈合，若停药后出现复发，需重新启动治疗。在笔者总结的104例PPV患儿中，有96例明确了预后情况，有8例（8.3%）仅采用局部GC即达到完全缓解，避免了系统使用GC带来的不良反应。

对局部治疗反应不佳的轻度和中重度PPV，首选GC系统治疗。中国PV诊疗指南中推荐GC（泼尼松）的初始剂量为0.5～1.25 mg/kg/d，通常不超过1.5 mg/kg/d[50]，欧美指南推荐剂量为0.5～1.5 mg/kg/d[46,51]，根据病情酌情调整激素用量。在笔者总结的104例PPV患儿中，有97例（93.2%）在治疗时系统使用了GC，其中74例（75.3%）使用了泼尼松，63例（85.1%）起始口服泼尼松剂量为0.5～1.5 mg/kg/d，有11例（14.9%）可能由于就诊时病情较为严重采用2 mg/kg/d的初始剂量。

对GC治疗反应不佳或不良反应严重的患儿，可考虑使用免疫抑制剂、生物制剂以及其他药物辅助治疗。在笔者总结的104例PPV患儿中，除GC外，使用率较高的药物主要是硫唑嘌呤（azathioprine，AZA）（24.0%）、氨苯砜（4，4’-diaminodiphenylsulfone，DDS）（21.7%）和利妥昔单抗（rituximab，RTX）（12.5%）。

AZA是治疗PV的一线免疫抑制剂，中国成人推荐剂量2～2.5 mg/kg/d（≤150 mg/d）[50]。欧美国家指南推荐剂量为1～3 mg/kg/d（100～200 mg/d）[46,51]。笔者总结的25例使用AZA治疗的PPV患儿中，有12例明确了剂量，为0.5～2 mg/kg/d（50～100 mg/d）。

DDS是一类具有抗炎特性的抗菌药物[52]，是治疗线状IgA病的一线用药，近几年的国内外指南暂未将其作为PV的一线用药。文献中对于儿童的推荐初始剂量为0.5 mg/kg/d，可逐渐增加至2 mg/kg/d，直至症状缓解[53]。

RTX是一种靶向作用于CD20阳性B细胞的单克隆抗体，已被建议作为中重度APV的一线疗法[51,54]，也被广泛用于治疗不同的儿童自身免疫性疾病[55-56]。RTX治疗APV的完全缓解率可达47.0%～89.5%[57]，但仅有少量研究报道了其在PPV中的应用。目前最广泛使用的两种输注RTX的方案是淋巴瘤方案（4次×375 mg/m^2，间隔1周，连续8周为一个疗程）和类风湿关节炎（rheumatoid arthritis，RA）方案（2次×1 000 mg，间隔2周，连续4周为一个疗程）[58]。中国成人推荐RA方案或在RA方案的基础上，将每次剂量降低为500 mg输注。欧洲皮肤性病学会推荐RA方案，美国皮肤性病学会则推荐淋巴瘤或RA方案。采用RTX治疗的患儿中，大部分使用淋巴瘤方案，有文献将淋巴瘤方案中输注间隔时间改为2 w[59]。就疗效和安全性而言，越来越多证据支持RTX作为难治性及GC不敏感性PPV的替代治疗方案。

5.2　疾病管理

5.2.1　监测与随访　PPV患者可采用天疱疮疾病面积和活动指数（PDAI）或天疱疮严重度评分（ABSIS）等特定评分系统来定量评估疾病严重度和治疗前后的变化[60-61]。治疗开始时建议每2～4周进行一次随访，直至旧病损逐渐愈合，同时无新病损发生。病情基本稳定时建议每4～8周进行一次

随访，直至停药。停药后建议每8～16周进行一次随访，直到完全缓解并且血清抗Dsg抗体恢复正常[46]。建议在治疗开始时、治疗后3个月以及根据病情进展每3～6个月监测一次血清自身抗体水平，可选择ELISA（首选）或IIF[62-63]。

停药时机主要基于临床症状以及血清抗体检测水平，对于使用最低维持剂量后达到完全缓解，且抗Dsg抗体阴性的患者，在进行血浆促肾上腺皮质激素检查后可以考虑停止系统性GC治疗。APV的泼尼松减量方案暂未达成共识，中国与美国指南中推荐的泼尼松维持剂量为≤10 mg/d，欧洲指南中推荐的维持剂量为0.1 mg/kg。有文献建议在PPV治疗中，泼尼松长期维持剂量为5～20 mg/d[49]。笔者总结的104例PPV患儿中，有74例使用泼尼松治疗，维持剂量范围为隔日服药1.5～5.0 mg/d（隔日服药0.075～0.25 mg/kg）。因此，与成年人相比，PPV患者可考虑使用更低剂量维持。

5.2.2　GC不良反应的预防和处理　PPV患儿长期服用GC，可能会对生理功能和精神心理造成不良影响，更容易出现生长发育、代谢及眼部不良反应，例如生长发育延迟、骨质疏松、代谢性紊乱、肥胖、肾上腺抑制等[64-65]。在笔者总结的104例PPV患者中，有23例（22.1%）提及GC治疗后出现了严重不良反应。所有出现不良反应的患儿均伴有代谢相关不良反应，其中库欣样面容（73.9%）和体重增加（39.1%）最为常见，此外还包括水肿（4.3%）和高血压（17.4%）。7例（30.4%）患儿出现了生长与骨骼发育的不良反应，包括生长迟缓（17.4%）、骨质疏松（8.7%）和骨折（4.3%）。其他不良反应还包括口腔白色念珠菌感染（17.4%）、早期白内障（4.3%）及心理健康问题（4.3%）。

病情稳定时，尽可能减少口服GC的剂量和持续时间，预防骨质疏松和生长发育抑制。目前，尚未有药物被美国食品药品管理局（Food and Drug Administration，FDA）批准用于临床治疗GC导致儿童骨生长迟缓。对于需用药来改善生长情况的患儿，可考虑重组人生长激素、重组人胰岛素样生长因子-1等[66-67]。在决定是否对儿童骨质疏松进行药物干预时，需要考虑骨质疏松危险因素的可逆性和骨骼剩余的生长潜力，对于外源性因素诱导的骨质疏松，不一定需要药物治疗。研究表明，GC诱导的骨质疏松，在停药后骨量可能会自发恢复[68]。若停药后改善不明显，可考虑药物治疗，但由于证据有限，在儿童中使用缺乏相关指南。

患儿长期使用GC罹患白内障和青光眼的风险较成年人更高，因此应至少每6个月进行一次临床眼科评估。若出现无法通过药物或手术手段控制的眼压升高，建议降低剂量或停用GC，积极寻找其他替代药物[69-70]。

5.2.3　其他药物不良反应的预防和处理　儿童使用免疫抑制剂常见的副作用包括肾毒性、高血压、高血糖、骨病、骨髓抑制、肿瘤和感染等[71]。尽管在笔者总结的41例使用免疫抑制剂治疗的PPV患儿中，未见严重不良反应的发生，但在PPV治疗中，免疫抑制剂的选择仍需慎重，且必须严格监控其不良反应。

除以上常规不良反应，AZA使用还可能造成缺乏巯基嘌呤甲基转移酶活性的患者出现严重的骨髓抑制，建议使用前进行TPMT及NUDT15基因型检测[72-73]。RTX主要不良反应包括：输注反应、过敏反应、感染、乙型肝炎病毒再激活等[54]。建议患者首次输注之后前3个月至少每月进行1次实验室检查，后续至少每3个月复查1次[46,50-51]。

5.2.4　疫苗接种　美国传染病学会建议PPV患儿与健康儿童按照相同的时间表进行常规灭活疫苗接种。尽可能在服用免疫抑制药物前2周接种疫苗。免疫抑制患者禁用活疫苗（带状疱疹疫苗、活流感疫苗、麻疹腮腺炎-风疹疫苗）。若有特殊需要，建议在开始免疫抑制治疗前4周接种活疫苗[74]。

6　预后特点

自GC出现后，天疱疮患者（尤其是PV）死亡率从75%降低到30%，免疫抑制剂的辅助应用使得疾病本身所致的死亡率进一步下降至5%～30%[11]。常见的死亡原因为皮肤和黏膜剥离导致的体液和蛋白质流失，以及大剂量使用GC和免疫抑制剂导致的感染等并发症[75]。

PV的缓解率与随访时间和治疗方式有较大关系。Cholera等[76]汇总了1 924例初诊年龄4～89岁的PV患者治疗数据，随访时间从9个月至22年。其中，321例（16.7%）患者完全缓解（停止治疗），894例（46.5%）患者部分缓解（低剂量维持治疗），485例（25.2%）患者仍在接受治疗，177例（9.2%）患者死亡，47例（2.4%）患者被归类为无反应并转诊到其他地方治疗。此外，研究显示天疱疮完全缓解率为12%～57.1%，部分缓解率为19.3%～62.3%[77-78]。笔者总结的104例PPV患儿随访时间

为1～120个月，平均38个月。PPV患儿的预后优于APV患者，在88例明确预后情况的患儿中，有41例（46.6%）在经过治疗后完全缓解（停止治疗），有36例（40.9%）部分缓解（低剂量维持），有10例患儿仍在接受治疗（11.4%），仅有1例（1.1%）患儿使用激素后出现严重的并发症，最终死于肺炎和败血症[79]。PPV患儿与APV患者死亡率差异的原因可能是由于成人患者需要使用更长时间的GC和免疫抑制剂才能达到减量要求，会导致严重的免疫抑制。同时，成年人往往患有更多的基础疾病，这一定程度上增加了死亡的风险。

7　总结与展望

PPV的病因、临床特点和组织病理学特点与APV具有一定的相似性，关键在于早期诊断，及时采取正确的治疗手段。由于该病较为罕见，且儿童患者表达能力、依从性相对较差，诊断比成人患者更为困难，表1总结了APV与PPV的疾病特征对比。口腔黏膜是PPV最易累及的部位，口腔医师应保持警惕，对于原因不明的口腔黏膜水疱、糜烂及溃疡，应考虑PPV的可能。患儿的依从性是影响PPV治疗效果的重要因素，对PPV进行临床管理时，应注意对患儿和家长进行健康教育，充分发挥家庭的支持、鼓励、监督和促进作用。PPV可能累及多个器官，口腔黏膜科医师除关注口腔病损外，还应关注患儿全身其他系统的症状和体征，必要时通过多学科团队合作及时进行干预，保护患儿的身心健康。

表1　APV与PPV的疾病特征对比

Table 1　Comparison of disease characteristics between APV and PPV

		PPV	APV
Age of onset		≤12 years old　29.8% 12~18 years old　70.2%	>18 years old (Onset typically occurs 40–60 years old)
Gender		Male　51.0% Female　49.0%	The ratio of male to female is 1:1.1
Lesion characteristics	Oral mucosa	87.3%	71.4%–98.1%
	Lips	25.0%	22.0%–46.2%
	Buccal mucosa	27.9%	30.0%–63.2%
	Tongue	19.2%	22.0%–45.3%
	Palate	15.4%	20.0%
	Floor of the mouth	2.9%	8.0%
	Gingiva	19.2%	28.0%
	Other mucosa	52.9%	20.0%–50.0%
	Nasal mucosa	5.8%	16.0%
	Pharynx and throat	16.3%	44.0%–55.0%
	Esophagus	3.8%	3.6%
	Conjunctiva	9.6%	2.0%–16.0%
	Genitalia	28.8%	3.0%
	Perianal area	6.7%	2.0%
	Skin	80.4%	16.0%–68.4%
	Scalp	17.3%	6.0%
	Face	19.2%	28.0%
	Limbs	39.4%	14.0%
	Trunk	51.0%	20.0%
	Fingernails/toenails	3.8%	13.0%
	Mucous membrane only	19.6%	18.0%
	Skin only	8.8%	11.6%
	Skin+ Mucous membrane	71.6%	70.4%

Laboratory examination	HE	98.0%	77.8%-100.0%
	DIF	98.9%	60.0%-100.0%
	IIF	91.4%	70.0%-90.0%
	Antibodies to Dsg1(ELISA)	73.7%	46.0%-75.0%
	Antibodies to Dsg3(ELISA)	100.0%	90.0%
Therapeutic regimen	Topical drug	7.7%	26.0%-39.0%
	Systemic administration		
	GC		
	Prednisone	71.2%	66.0%
	Others	22.1%	34.0%
	Immunosuppressor		
	AZA	24.0%	8.0%
	MMF	6.7%	18.0%
	CTX	3.8%	2.0%
	MTX	2.9%	2.0%
	CsA	1.9%	2.0%
	Biologicals		
	RTX	12.5%	8.0%
	Other drugs		
	DDS	21.7%	4.0%
Adverse reaction of GC	Growth and bone development	Growth retardation (17.4%), osteoporosis (8.7%), abnormal bone development, fracture (4.3%)	The risk of osteoporosis and fracture is increased
	Metabolism	Cushingoid face (73.9%), significant weight gain or obesity (39.1%), impaired glucose tolerance or diabetes, increased susceptibility to edema (4.3%) or hypertension (17.4%), affecting sex hormone levels (irregular menstruation and abnormal sexual development)	Cushing's appearance, weight gain or obesity, insulin resistance or diabetes, edema or hypertension, affected sex hormone levels (irregular menstruation, hirsutism, decreased testicular function)
	Immunosuppression and infection	Increased risk of infection (17.4%) and decreased vaccine response	Increased risk of infection
	Complication of eyes	More sensitive, especially increased eye pressure, cataracts (4.3%), and eye infections	Progressive and occult, and more closely related to systemic diseases
	Psychological health	Mood swings, anxiety, depression, and learning problems (4.3%)	Depression, anxiety, and changes in cognitive function
Mortality		1.1%	5.0%-30.0%
Remission rate	Complete response rate(discontinuation)	46.6%	16.7%
	Partial response rate (low dose maintenance)	40.9%	46.5%

References in Table 1: [8-11, 30-35, 39-41, 45-46, 48, 50-51, 76-78]. PPV: pediatric pemphigus vulgaris; APV: adult pemphigus vulgaris; HE: histopathological examination; DIF: Direct immunofluorescence; IIF: indirect immunofluorescence; ELISA: enzyme-linked immunosorbent assay; Dsg: desmoglein glycoprotein; GC: glucocorticoid; AZA: azathioprine; MMF: mortemycophenolate; CTX: cyclophosphamide; MTX: methotrexate; CsA: cyclosporine A; RTX: rituximab; DDS: 4, 4'-diaminodiphenylsulfone

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