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Diagnostic Criteria in Dermatology

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ABSTRACT

Diagnostic criteria are defined as group of features that can collectively be used to diagnose a condition. Diagnostic criteria are used for diagnosing those skin conditions that have heterogenous presentation and do not have a single specific clinical, laboratory or pathological feature.

Keywords: Diagnostic criteria, Atopic Dermatitis, Sweets Syndrome, Pyoderma Gangrenosum

INTRODUCTION

Diagnostic criteria are defined as group of features that can collectively be used to diagnose a condition. Diagnostic criteria are used for diagnosing those skin conditions that have heterogenous presentation and do not have a single specific clinical, laboratory or pathological feature. With improved knowledge of disease pathogenesis and advancement in diagnostic tools diagnostic criteria are updated accordingly to increase their specificity and sensitivity.

ATOPIC DERMATITIS

Atopic dermatitis is an inflammatory disease of skin with early onset consisting of eczematous lesions with itch. It follows the course of remissions and exacerbations, improves with age. Exact etiology of Atopic Dermatitis is unknown, environmental factors together with genetic factors (like filaggrin gene mutation) is believed to involved in pathogenesis. There are ten different diagnostic criteria used for diagnosing Atopic Dermatitis. The Hanifin and Rajka criteria is most commonly used, followed by the UK refinement of the Hanifin and Rajka criteria and Japanese Dermatological Association criteria.[1,2] Hanifin and Rajka modified and combined their diagnostic standard to propose new one (Table 1).

Table 1 Diagnostic Standard of Hanifin & Rajika.

1. Must have three or more basic features described below

1. Pruritus
2. Typical morphology and distribution
   - Flexural lichenification or linearity in adults
   - Facial and extensor eruptions in infants and children
3. Chronic or chronically relapsing dermatitis
4. Personal or family history of atopy (asthma, allergic rhinitis, atopic dermatitis)

2. Must have three or more following minor features:

1. Xerosis
2. Ichthyosis/palmar hyperlinearity/keratosis pilaris
3. Immediate (type I) skin test reaction
4. Elevated serum IgE
5. Early age of onset
6. Tendency toward cutaneous infections (especially *staph. aureus* and *herpes simplex*), impaired cell- mediated immunity

7. Tendency toward non-specific hand or foot dermatitis
8. Nipple eczema
9. Cheilitis
10. Recurrent conjunctivitis
11. Dennie-Morgan infraorbital fold
12. Keratoconus
13. Anterior subcapsular cataracts
14. Orbital darkening
15. Facial pallor, facial erythema
16. Pityriasis alba
17. Anterior neck folds
18. Itch when sweating
19. Intolerance to wool ad lipid solvents
20. Periofollicular accentuation
21. Food intolerance
22. Course influenced by environmental and emotional factors
23. White dermographism, delayed blanch

Infantile atopic dermatitis is difficult to diagnose even with Hanifin and Rajka criteria.

SWEET'S SYNDROME

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, was first described by Robert Douglas Sweet in 1964.[3] It presents as painful erythematous plaques, may be associated with fever, neutrophilia, leukocytosis. Dramatic response to systemic corticosteroids is characteristic of the disease.[4] Sweet's syndrome though primarily a dermatologic disorder is accompanied with features of systemic inflammation, including inflammation of the eye. Ocular manifestations include conjunctivitis, episcleritis, scleritis, limbal nodules, peripheral ulcerative keratitis, iritis, periorbital and orbital inflammation, dacryoadenitis, glaucoma, choroiditis, retinal vasculitis, and optic nerve involvement.[5,6,7] It can be associated with recurrent aseptic meningoencephalitis, known as Neuro-Sweet disease.[8]

Su and Liu proposed a set of major and minor criteria for the diagnosis of Sweet's Syndrome in 1986.[9] Von den Driesch modified these diagnostic criteria in 1994, according to which both major and two minor criteria were required for diagnosis of Sweet's Syndrome (Table 2).[10]

Table 2. Modified diagnostic criteria for Sweet's syndrome as proposed by von den Driesch

- **Major criteria**

1. Abrupt onset of tender or painful erythematous plaques or nodules, occasionally with vesicles, pustules, or blisters
2. Predominantly neutrophilic dermal infiltrate without leukocytoclastic vasculitis

- **Minor criteria**

1. Preceded by a nonspecific respiratory or gastrointestinal tract infection or vaccination or associated with:
   - Inflammatory diseases such as chronic autoimmune disorders, infections
   - Hemoproliferative disorders or solid malignant tumors
   - Pregnancy
2. Fever > 38°C
3. Abnormal laboratory values at presentation (three of four):
   - Erythrocyte sedimentation rate > 20 mm/h
   - Elevated C-reactive protein levels
   - Leukocytosis > 8,000
   - Neutrophilia > 70 %
4. Excellent response to treatment with systemic corticosteroids or potassium iodide

Walker and Cohen proposed separate diagnostic criteria for drug-induced Sweet's Syndrome in 1996 (Table 3).[11]

Table 3. Diagnostic criteria for drug-induced Sweet's syndrome*

- Abrupt onset of tender or painful erythematous plaques or nodules
- Dense dermal neutrophilic infiltrate *without* leukocytoclastic vasculitis

- Fever > 38°C
- Temporal relationship between drug ingestion and clinical presentation, OR temporally related recurrence after oral challenge
- Temporally related resolution of lesions following drug withdrawal or treatment with systemic corticosteroids

*All five criteria are required for the diagnosis of drug-induced SS.

HENOCH SCHONLEIN PURPURA

Henoch-Schönlein purpura is an IgA-mediated immune vasculitis that primarily involves the small vessels of the joints, kidneys, gastrointestinal (GI) tract, skin and less commonly, the central nervous system and lungs. There is no definitive tests for diagnosis of HSP.

The European League Against Rheumatism, the Paediatric Rheumatology International Trials Organization and the Paediatric Rheumatology European Society classification criteria 2010 is an advancement over older American College of Rheumatology (ACR) classification criteria for HSP. [12,13]

Table 4. EULAR/PRINTO/PRES Diagnostic criteria for Henoch-Schönlein purpura (HSP):

Mandatory criterion: Purpura (commonly palpable and in crops) or petechiae with lower limb predominance (not related to thrombocytopenia)

Minimum 1 out of 4 criteria

1. Diffuse colicky abdominal pain with acute onset
2. Histopathology showing leukocytoclastic vasculitis or proliferative glomerulonephritis, with predominant immunoglobulin A (IgA) deposits
3. Arthritis or arthralgia of acute onset*
4. Renal involvement in the form of proteinuria (>0.3g/24h or >30 mmol/mg of urine albumin/creatinine ratio) or haematuria (blood cell casts in urinary sediment or ≥2+ on dipstick)

*in acute arthralgia there is no joint swelling or limitation of motion, unlike acute arthritis

BEHCET'S DISEASE

Behçet disease (BD) is an multisystemic, inflammatory vasculopathy, first described by Hulusi Behcet in 1931. It has a relapsing-remitting course with oral apthae, genital ulcerations and iritis considered hallmarks of the disease.[14] BD was first described as a dermatologic disease, however, ocular, cardiovascular, articular, neurological, and gastrointestinal manifestations are also common.[15,16,17] While the exact etiology and pathogenesis is unknown, there is a strong correlation with human leukocyte antigens, specifically HLA-B51.

The International Criteria for Behcet's Disease (ICBD) were presented to the International Conference of Behcet's Disease in 2006. (Table 5).

Table 5. International Criteria for Behcet's Disease

Sign/Symptom	Points
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular manifestations	1
Positive pathergy test[a]	1

1. [a] Pathergy test is optional. Where a pathergy test is conducted, 1 extra point may be added for a positive result. Adapted from International Team for the Revision of the International Criteria for Behçet's Disease (ITR-ICBD).
2. Patients with a score of < 3 were considered as not having BD. A score of 3 was considered probable BD, and a score of ≥4 indicated a definitive diagnosis of BD.

PYODERMA GANGRENOSUM

Pyoderma gangrenosum (PG) is a reactive non-infectious serious ulcerating skin disease, falling under the spectrum of the neutrophilic dermatoses. Several variants exist, which lead to delay in diagnosis. Typical presentation is extremely painful erythematous lesion progressing rapidly to a blistered or necrotic ulcer with ragged undermined edge and a violaceous/erythematous border.[18] The lower legs are most frequently affected site in PG. The lesion may develop at the sites of minor trauma, a phenomenon known as 'pathergy'.[19]

There is no validated, established diagnostic clinical or pathological criteria to diagnose PG. Su *et al* have proposed a diagnostic tool for diagnosing PG (Table 6).[20]

Table 6. Su et al diagnostic tool for PG.

Major criteria Rapid progression of a painful necrolytic cutaneous ulcer with an irregular, violaceous and undermined border Other causes of cutaneous ulceration excluded
Minor criteria History of pathergy or cribriform scarring clinically Associated systemic disease (inflammatory bowel disease, arthritis, IgA gammopathy or underlying malignancy) Classic histopathological findings Treatment response (rapid response to systemic steroid treatment – 50% improvement in 1 month) Diagnosis requires 2 major and 2 minor criteria.

Maverakis *et al* have proposed new criteria, yet to be widely adopted (Table 6).[21]

Table 6. Improved diagnostic tool for Pyoderma Gangrenosum.

Major criteria Biopsy of ulcer edge demonstrating neutrophilic infiltrate
Minor criteria Exclusion of infection Pathergy History of inflammatory bowel disease or inflammatory arthritis History of papule, pustule or vesicle ulcerating within 4 days of appearing Peripheral erythema, undermining border, and tenderness at ulceration site Multiple ulcerations, at least one on anterior lower leg Cribriform or 'wrinkled paper' scar(s) at healed ulcer sites Decreased ulcer size within 1 month of initiating immunosuppressive medication(s)

Diagnosis requires one major and four minor criteria

Declaration by Authors

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REFERENCE

1. Hanifin, J.M. and Rajka, G.: Diagnostic features of atopic dermatitis. A*cta Derm Venereol Suppl (Stockh)* 1980; 92: 44–47.
2. Vakharia, P. P., Chopra, R. & Silverberg, J. I. Systematic review of diagnostic criteria used in atopic dermatitis randomized controlled trials. Am. J. Clin. Dermatol. 19, 15–22 (2018)
3. Aggarwal R, Ringold S, Khanna D, et al. Distinctions between diagnostic and classification criteria? *Arthritis Care Res.* (2015) 67:891–7. doi: 10.1002/acr.22583
4. Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol* 1964; 76: 349–56.
5. Rochet NM, Chavan RN, Cappel MA et al. Sweet syndrome: Clinical presentation, associations, and response to treatment in 77 patients. *J Am Acad Dermatol* 2013; 69: 557– 64.
6. Gottlieb CC, Mishra A, Belliveau D, et al. Ocular involvement in acute febrile neutrophilic dermatosis (Sweet syndrome): new cases and review of the literature. *Surv Ophthalmol*. 2008;53(3):219-226. doi:10.1016/j.survophthal.2008.02.006
7. Marcoval J, Martín-Callizo C, Valentí-Medina F, et al. Sweet syndrome: long-term follow-up of 138 patients. *Clinical and Experimental Dermatology*. 2016; 41(7):741-746. doi:10.1111/ced.12899
8. Baartman B, Kosari P, Warren CC, et al. Sight-threatening ocular manifestations of acute febrile neutrophilic dermatosis

(Sweet's syndrome). *Dermatology*. 2014; 228(3): 193-197. doi:10.1159/000357729
9. Hisanaga K, Iwasaki Y, Itoyama Y, Neuro-Sweet Disease Study Group. Neuro-Sweet disease: clinical manifestations and criteria for diagnosis. *Neurology*. 2005; 64(10): 1756-1761. doi:10.1212/01.WNL.0000161848.34159.B5
10. Su WPD, Liu HN. Diagnostic criteria for Sweet's syndrome. *Cutis* 1986; 37: 167–74.
11. Von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol*1994; 31: 535– 56.
12. Walker DC, Cohen PR. Trimethoprim-sulfamethoxazole associated acute febrile neutrophilic dermatosis: case report and review of drug-induced Sweet's syndrome. *J Am Acad Dermatol* 1996; 34: 918– 23.
13. Mills JA, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schonlein purpura. Arthritis Rheum. 1990;33(8):1114–21.
14. Ozen S, Pistorio A, Iusan SM, et al. EULAR/PRINTO/PRES criteria for Henoch-Schonlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. Ann Rheum Dis. 2010;69(5):798–806.
15. Yazici H, Seyahi E, Hatemi G, et al. Behçet syndrome: a contemporary view. Nat Rev Rheumatol. 2018;14(2):107.
16. Davatchi F, Chams-Davatchi C, Shams H, et al. Behcet's disease: epidemiology, clinical manifestations, and diagnosis. Expert Rev Clin Immunol. 2017;13(1):57–65.
17. Hatemi I, Hatemi G, Celik AF. Gastrointestinal involvement in Behçet disease. Rheum Dis Clin N Am. 2018; 44(1):45–64.
18. Krause I, Weinberger A. Behcet's disease. Curr Opin Rheumatol. 2008;20(1):82–7.
19. Sassolas B, Le Ru Y, Plantin P, et al. Pyoderma gangrenosum with pathergic phenomenon in pregnancy. *Br J Dermatol* 2000;142:827–8
20. Su WP, Davis MD, Weenig RH, et al Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. *Int J Dermatol* 2004;43:790–800.
21. Maverakis E, Ma C, Shinkai K, et al. Diagnostic criteria of ulcerative pyoderma gangrenosum. A Delphi consensus of international experts. *JAMA Dermatol* 2018;154:461–6.

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