

ii18 REPORT Psoriasis: epidemiology, clinical features, and quality of life R G B
 Langley, G G Krueger, C E M Griffiths Ann Rheum Dis 2005;64(Suppl III):ii18–ii23 doi:10
 1136/ard.2004.033217 The molecular genetic basis of psoriasis is complex with
 Psoriasis is a common chronic, recurrent, immunemediated evidence that multiple genes are
 involved Seven major disease of the skin and joints It can have a significant psoriasis
 susceptibility loci have been reported Many negative impact on the physical, emotional,
 and, psycho- investigators have established that a major susceptibility social **wellbeing**
 of affected patients Psoriasis is found locus for psoriasis is at 6p21, referred to as
 PSORS4 and is worldwide but the prevalence varies among different ethnic overrepresented
 in all populations tested 10–15 As noted, an groups
 It has a strong genetic component but environmental association between psoriasis and other
 loci has also been factors such as infections can play an important role in the
 reported on chromosomes 1p (PSORS7), 141q (PSORS4), 16p (PSORS1), 17q (PSORS3), 1817q (PSORS2), 19 and 19p
 There are several clinical cutaneous (PSORS5), 174q (PSORS3), 1817q (PSORS2), 19 and 19p
 manifestations of psoriasis but most commonly the disease (PSORS6)
 20 The strength of associations between such genes presents as chronic, symmetrical,
 erythematous, scaling and susceptibility to psoriasis, apart from PSORS1, is variable papules
 and plaques The epidemiology, clinical features, as replication of these findings has
 been incomplete The and impact on quality of life of psoriasis are reviewed difficulty of
 confirming psoriasis susceptibility loci may
 relate, in part, to heterogeneity among different populations
 Whereas the existence of a genetic component in psoriasis is
 certain, the exact location of the genes involved remain to
 This paper reviews the epidemiology and clinical features be definitely determined
 of psoriasis and its impact on patients' quality of life CLINICAL FEATURES EPIDEMIOLOGY Psoriasis
 is a **papulosquamous** disease with variable mor- Although psoriasis occurs worldwide, its
 prevalence varies phology, distribution, severity, and course Papulosquamous
 considerably In the USA, approximately 2% of the popula- diseases are characterised by
 scaling papules (raised lesions tion is affected High rates of psoriasis have been reported in
 , 1 cm in diameter) and plaques (raised **lesions** 1 cm in people of the Faroe islands, where
 one study found 2.8% of diameter) Other **papulosquamous** diseases that maybe
 the population to be affected 1 The prevalence of psoriasis is considered in the differential
 diagnosis include tinea infec- low in certain ethnic groups such as the Japanese, and maytions,
pityriasis rosea, and **lichen planus** The lesions of
 be absent in aboriginal Australians² and Indians from South psoriasis are distinct from these
 other entities and are America³ classically very well circumscribed, circular, red
 papules or Psoriasis can present at any age and has been reported at plaques with a grey or
 silvery-white, dry scale In addition, birth and in older people of advanced age Accurate
 the lesions are typically distributed symmetrically on the
 determination of the age of onset of psoriasis is problematic,
 scalp, elbows, knees, lumbosacral area, and in the body folds as studies which do so typically
 rely on a patient's recall of (fig 1) Psoriasis may also develop at the site of trauma
 or the onset of lesions or determine the onset from the injury, known as Koebner's
 phenomenon If psoriasis is physician's diagnosis as recorded on the initial visit Data
 progressive or uncontrolled, it can result in a generalised
 based on patient recall can be inaccurate; determining onset exfoliative **erythroderma** **Nail**
involvement maybe present, based on first visit to a physician could underestimate the
 particularly if psoriatic arthritis (PsA) is present time of disease occurrence, as minimal
 disease maybe Occasionally psoriasis may involve the oral mucosa or the present for years
 before a consultation is sought A bimodal tongue
 When the tongue is involved, the dorsal surface may

age of onset has been recognised in several large studies. The mean age of onset for the first presentation of psoriasis can range from 15 to 20 years of age, with a second peak on a daily basis, can assume distinct annular patterns and occurring at 55–60 years. 4–7 may resemble a map, hence the term geographic tongue.

Henseler and Christophers examined a series of 2147 Psoriasis patients and reported two clinical presentations of psoriasis, Type I and Type II, distinguished by age at onset and severity. Despitethe classic presentation described by Landells, Type I begins on or before age 40 years; Type II begins after the age of 40 years. Above, the morphology can range from small tear shaped of 40 years. Type I disease accounts for more than 75% of papules (guttate psoriasis) to pustules (pustular psoriasis) cases.

7 Patients with early onset, or type I psoriasis, tended to have more severe disease than those with late onset (erythrodermic psoriasis). In addition, these different forms of psoriasis may be localised or widespread and disabling. Further, psoriasis may have strong associations with human leucocyte antigen (HLA)-Cw6 in patients with early onset, compared with late onset of psoriasis. The course and widespread involvement of psoriasis is unpredictable. In one study, 39% of patients complaining of intense pruritus or burning. The patients reported complete remission of disease for between one and 54 years. 8 Higher figures have been reported in Abbreviations: PASI, Psoriasis Area and Severity Index; PsA, psoriatic arthritis; PSI, Salford Psoriasis Index; RA, rheumatoid arthritis. www.nnrheumdis.com

Psoriasis: epidemiology, clinical features, and quality of life. ii19 Figure 2

Nummular (coin-sized) lesions of psoriasis. 2–10 mm diameter lesions of psoriasis. These are usually distributed in a centripetal fashion although guttate lesions can also involve the head and limbs. Classically, guttate psoriasis occurs shortly after an acute group B streptococcal infection of the pharynx or tonsils and can be elbows the presenting episode of psoriasis in children or, occasionally, adults. The number of lesions may range from five or 10 various types and presentations of psoriasis are outlined to over 100. Guttate psoriasis accounts for 2% of the total below cases of psoriasis. In children, an acute episode of guttate psoriasis is usually self-limiting; in adults, guttate flares may complicate chronic plaque disease. Although few studies have assessed the long term prognosis of children with acute plaque psoriasis, one small study revealed that 33% of patients with acute guttate psoriasis eventually developed chronic plaque disease. 21 nummular (coin-sized) plaques (fig 2). The lesions may initially begin as erythematous macules (flat and, 1 cm) or papules, extend peripherally, and coalesce to form plaques of several centimetres in diameter. A white blanching Psoriasis affecting the flexures, particularly inframammary, ring, known as Woronoff's ring, may be observed in the skin perineal, and axillary, is distinct morphologically from surrounding a psoriatic plaque. With gradual peripheral extension, plaques may develop different configurations. Flexural lesions are devoid of scale and appear as red, shiny, well demarcated plaques occasionally confused with candida, intertrigo, and dermatophyte infections. psoriasis gyrata—in

which curved linear patterns predominate. **Erythroderma annular** psoriasis—in which ring-like lesions develop. Total or subtotal involvement of the skin by active psoriasis is secondary to central clearing. It is known as erythroderma and may take one of two forms: psoriasis **follicularis**—in which minute scaly papules are. Firstly, chronic plaque psoriasis may gradually progress as present at the openings of pilosebaceous follicles. Plaques become confluent and extensive. Secondly, erythro- The terms rupoid and ostraceous relate to distinct morphologies. Erythroderma may be a manifestation of unstable psoriasis. Guttate psoriasis is a subtype of plaque psoriasis. Rupoid plaques are small, precipitated by infection, tar, drugs, or withdrawal of (2–5 cm in diameter) and highly hyperkeratotic, resembling corticosteroids. Erythroderma may impair the thermoregulatory capacity of the skin, leading to hypothermia, high output heart failure, and metabolic changes including hypoalbuminaemia, and anaemia due to loss of iron, vitamin B₁₂, and folate. Scale is typically present in psoriasis, is characteristically silvery white, and can vary in thickness. Removal of scale may reveal tiny bleeding points (Auspitz sign). The amount of scale varies among patients and even at different sites on the body. Generalised **pustular psoriasis** (von Zumbusch) is rare and a given patient represents active, unstable disease. Precipitants include psoriasis, scaling can be minimal and erythema may be the withdrawal of systemic or potent topical corticosteroids and predominant clinical signs are infections. The patient is pyrexial, with red, painful, inflamed skin studded with monomorphic, sterile pustules, which may coalesce to form sheets. Patients with generalised pustular psoriasis, from the Greek word gutta meaning a drop, frequently need to be admitted to the hospital. Langley, Krueger, Griffiths describes the **acute onset** of a myriad of small, management. **Palmoplantar pustulosis** presents as sterile, yellow pustules on a background of erythema and scaling affecting the palms and/or soles (fig 3). The pustules are tender and fade to form dark brown coloration with adherent scale/crust. **Palmoplantar pustulosis** is frequently associated with psoriatic nail involvement. Approximately 25% of cases are associated with classic **psoriasis vulgaris**, but it is now believed that **palmoplantar pustulosis** may not be a form of psoriasis. This conclusion is derived from genetic studies showing no association with HLA-Cw6 or other markers on chromosome 6p—which are linked to **chronic plaque** and guttate psoriasis. The demographics of palmoplantar pustulosis are markedly different from those of chronic plaque psoriasis in that it more commonly affects women (9:1), presents most commonly between the ages of 40 and 60 years, and has a very striking association with smoking, either current or past, in up to 95% of subjects. **Psoriatic nail disease** Finger nails are more commonly affected than toenails. The commonest finding is small pits in the **nail plate**, resulting from defective nail formation in the proximal portion of the nail matrix (fig 4). The nail may also detach from the bed at its distal or lateral attachments, known as onycholysis (see dystrophic, and show orange-yellow areas (oil spots) (fig 4). Orange-yellow areas may be present beneath the nail plate and are termed “oil spots”. In addition, the **nail plate** may become thickened, dystrophic, and discolored (fig 5) leading to everyday **disability** leading to depression and suicidal ideas. Yellow, keratinous material may collect under the nail plate in more than 5% of patients. **Psoriasis** is well known as a subungual hyperkeratosis. Recent work has identified that pathological worry and anxiety occur in at least a third of patients with psoriasis and significantly affect the quality of life and psychological well-being.

ASPECTS of psychological interpersonal difficulties impinge on all OF PSORIASIS aspects of the patient's daily life 2931 The two main Although psoriasis generally does not affect survival, it contributor to stress in patients with psoriasis are engaging certainly has a number of major negative effects on patients, in avoidance behaviour and the belief that they are being demonstrable by a significant detriment to quality of life 24 evaluated on the basis of their skin disease This constraining, Despite this, most clinical trials of new treatments for avoidance behaviour may lead to low grade persistent stress psoriasis focus on "objective" physical measures for the Intriguingly, there is no significant relation between either primary endpoint of efficacy This is incongruous as it is the the physical severity or anatomic location of psoriasis and improvement in quality of life that patients and physicians **psychological disability** 3233 This observation implies that rely upon when selecting treatment Impairment of quality of "severity" of psoriasis is a composite of physical and life has been highlighted particularly by the work of psychological factors, a disparity further highlighted by the Finlay 2526 Patients with psoriasis have a reduction in their Psoriasis Disability Index 34 Stress in the form of pathological quality of life similar to or worse than patients with other worry has a deleterious effect on response to therapy For chronic diseases, such as ischaemic **heart disease** and instance, in patients undergoing PUVA therapy, those who diabetes 25 That patients with psoriasis feel stigmatised by are delineated as being high or pathological worriers clear the condition is well established 30 This of itself contributes to significantly more slowly, if at all, as compared with their www.nnrheumdis.com Psoriasis: epidemiology, clinical features, and quality of life ii21 N counterparts who are low worriers 35 Psychological intervention may play a role in the management of psoriasis, the PASI N particularly in the form of cognitive behavioural stress P—Psychosocial **disability**: measured as 0–10 on a visual management 86 This form of intervention, when used as an analogue scale N adjunct to regular pharmacological therapy, produces a I—Interventions: a cumulative historical record of sys- significant additional benefit identified as improvement in temic therapies, episodes of erythroderma, etc clinical severity of **disease** How **psychological distress** exacerbates or triggers psoriasis is poorly understood Up to The SPl is represented as three figures such as 9, 7, 6 and is a 60% of patients describe stress as being a key "exacerbator" guide to the difficulty of treating any one patient at a certain or trigger of their disease 83738 It is known that psychological time stress has the potential to regulate the immune response, and Physicians evaluating **chronic disease** states, such as RA there is emerging evidence that abnormal neuroendocrine and inflammatory bowel disease (IBD), have used quality of responses to stress may contribute to the pathogenesis of life data to assess treatment efficacy The Inflammatory chronic autoimmune **diseases**, as has been described for Bowel Disease Questionnaire, a commonly used quality of life **rheumatoid arthritis (RA)** 39 It is likely that, in some patients measure for IBD, has been validated in Crohn's disease 48 and with psoriasis, there is an abnormal hypothalamic–adrenal has been shown to correlate highly with the commonly used axis response to acute stress, undoubtedly an area deserving objective measure, the Crohn's Disease Activity Index of further investigation (CDAI) 49 The CDAI also incorporates a quality of life Many instruments have been generated to measure aspects assessment, "the patient's sense of wellbeing", as one of of disease on quality of life Some reflect general health the eight measurable items 50 In the American College of status, some reflect on skin disease in general, and yet others Rheumatology (ACR) improvement criteria for RA, a quality

assess the impact of psoriasis and PsA (table 1). The current of life measure is often employed as the measure of metrics for quality of life in psoriasis generally measure on disability 51. Moreover, ACR response rates have been found in two categories, the physical aspects of disease (pain, itch, to be higher when quality of life criteria are used instead of etc) or the mental aspects of disease (self perception, objective measures, such as grip strength, to assess physical interaction with others, etc). To have a maximal quality of function/disability 52 life, one needs to be able to participate in all aspects of life. For psoriasis, many quality of life instruments have been including effective interaction with others and carrying out developed and tested in clinical trials to assess treatment physical responsibilities, both at work and at home. **Patient response** where the primary endpoint is the number of oriented quality of life measures are particularly beneficial in patients gaining a 75% reduction in the Psoriasis Area and chronic diseases as they assess how the disease affects a Severity Index (PASI) relative to placebo. Table 1 lists these persons socially, psychologically, and physically 47 and a few elements of each. In a review of trials where both Furthermore, quality of life measures take into account the physical measures and quality of life were collected, two effects of the treatment on the patient. **Quality of life** data things stood out. First, the correlation with the physical fulfils the role of measuring the intangible changes in a measure, such as the PASI, and quality of life is generally patient's life that determine "treatment success". For a very poor, the correlation coefficient being less than 0.2 clinically meaningful change to exist for psoriasis and other. Second, the improvement in quality of life over time generally chronic, non-life **threatening** diseases, a treatment must parallel the physical measure 53. This supports the notion that provide an improvement in the patient's quality of life. In an quality of life and the PASI measure two different aspects of attempt to provide an holistic assessment of overall disease. Given that it is the promise of change in quality of severity, a specific tool has been developed—the Salford life by a given treatment that patients and physician rely on. Psoriasis Index (SPI) 32: in choosing treatment, it is not surprising that considerable Table 1.

Instruments used in assessing quality of life in psoriasis and psoriatic arthritis	Name
Abbreviation Features Medical Outcomes Study 36 Item Short Form 36	SF-36
36 items; eight scales for physical and mental health; used to compare quality of life of skin disease with other disease	Nottingham Health Profile 40
NHP	
38 items; six scales ranging from physical mobility to socialisation	Sickness Impact Profile 41
SIP	
136 items; 12 scales for physical and mental health, as well as sleep, eating, work, recreation, etc	12 Item General Health Questionnaire 42
GHQ	
12 item questionnaire with higher correlation to PASI than others	Dermatology Life Quality Index 43
DLQI	
Widely used; 12 reports, 2500 patients, of use in psoriasis; is internally consistent, correlates with other quality of life tools—for example PSORIQoL, PQoLI, PDI, etc	Psoriasis Disability Index 44
PDI	
15 questions for functional disability in psoriasis	Skindex-29 42
Skindex-29	
29 items and scales derived from Skindex-61	Impact of Psoriasis Questionnaire 42
IPSQ	
Assesses psychosocial impact of psoriasis	Psoriasis Life Stress Inventory 42
PLSI	
Stress inventory that correlates with DLQI and IPSQ	Dermatology Specific Quality of Life Instrument 40
DSQLI	
52 items, eight global items (physical symptoms to appearance and severity); seven scales	Dermatology Quality of Life Scales 40
DQLS	
41 items with three scales	Psoriatic Arthritis Specific Measure
PsAQoL	
Series of questions generated from interviews with subjects with psoriatic arthritis, of Quality of Life 45	narrowed from 51 to 20 with Rasch Analysis; validated and reliable
Psoriasis Specific Measure of Quality of Life 46	PSORIQoL
Series of questions generated from interviews with psoriatic subjects, narrowed from	

61 to 25 using Rasch Analysis of each round of questioning Psoriasis Quality of Life Questionnaire 46
 PQoL fashioned after DQoL with specificity for psoriasis Dermatology Utilities 47 DU
 Quality of life instruments assess health status, DU are derived from decision theory
 and can be interpreted across diseases and populations www.nrrheumdis.com ii22
 Langley, Krueger, Griffiths thought and energy have gone into generating instruments 6
 Ferrandiz C, Pujol RM, Garcia-Patos V, Bordas X, Smandia JA Psoriasis of
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 A number of instruments have been designed to generate 7 Henseler T, Christophers E
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 house quality of life issues unique to that disease and hence 9 Yashuda T, Ishikawa E, Mori S
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 would be more robust in following disease specific quality of Cox AJ, eds Psoriasis
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 focused on Stanford, CA: Stanford University Press, 1971:25–34 generating a disease specific
 quality of life instrument by 10
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 extensive interview process
 evidence for further disease loci revealed by a two stage genome-wide search
 Following this, a Rasch analysis was used to select questions in psoriasis
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 This approach led to 25 and 20 question profiles that appear
 Evidence for two psoriasis susceptibility loci (HLA and 17q) and two novel
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 patients with psoriasis than those designed for general health 13
 Enlund F, Samuelsson L, Enerback C, Inerot A, Wahlstrom J, Yhr M, et al or specific for skin disease
 or psoriasis remains to be
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 Whereas the general health instruments, such as the SF-36 17q, but not to 4q Hum Hered 1999;49:2–8 4
 Veal CD, Clough RL, Barber RC, Mason S, Tillman D, Ferry B, et al
 (see table 1), can be used to compare the burden of disease of
 Identification of a novel psoriasis susceptibility locus at 1p and evidence of different diseases
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 J Med Genet 2001;38:7–13 Instruments are not good at incorporating outcomes into 15
 Nair RP, Stuart P, Henseler T, Jenisch S, Chian NV, Westphal E, et al
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patients to indicate their willingness to trade disease free status for the remainder of 17 Enlund F, Samuelsson L, Enerback C, Inerot A, Wahlstrom J, Yhr M, et al their lives in exchange for a reduction in their lifespan and to Psoriasis susceptibility locus in chromosome region 3q21 identified in patients from southwest Sweden Eur J Hum Genet 1999;7:783–90

Indicate the amount of reduction they would be willing to 18 Matthews D, Fry L, Powles A, Weber J, McCarthy M, Fisher E, et al Evidence accept As an example, patients in follow up for psoriasis that a locus for familial psoriasis maps to chromosome 4q Nat Genet indicated a willingness to trade 28 years of their remaining 1996;14:231–3

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CONCLUSION 21 Martin BA, Chalmers RJ, Telfer NR How great is the risk of further psoriasis At this time, there are many instruments to measure quality following a single episode of acute guttate psoriasis? Arch Dermatol 1996;132:717–18

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