Review Article

Pustular Psoriasis in Children- a Review

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Abstract

Pustular psoriasis is a rare form of psoriasis in the pediatric population with the acute generalized and annular variants being the most common. There are two groups of pustular psoriasis – one with a history of psoriasis vulgaris and the other without a history of psoriasis vulgaris which differ in various aspects including the age at onset and triggering factors. Several triggering factors have been implicated in generalized pustular psoriasis, the removal or treatment of which can allow the process to settle, which include upper respiratory tract and urinary tract infections, abrupt cessation of oral steroids and cyclosporine, sunburn, tar therapy, hypocalcemia and vaccination. But, generalized pustular psoriasis may present with potential life threatening complications warranting aggressive approach with various treatment modalities like retinoids, methotrexate, cyclosporine, dapsone and biologics which are frequently being used in children. However, the management issues in the pediatric age group are challenging pertaining to a host of precipitating factors, limited clinical experience with the optimal use of these agents in children, long term safety profile of these agents in the long run in children and the lack of long term follow up studies.

Key words: Pustular psoriasis; children; complications; retinoids; methotrexate; cyclosporine; dapsone; biologics

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Introduction

Psoriasis vulgaris in childhood is a well-recognized disease contributing to about 4% of the childhood dermatoses.[1] Though all clinical types of psoriasis occur in children, pustular and erythrodermic psoriasis are rare and pustular psoriasis is the most severe variant of childhood psoriasis.[2]

Classification of Pustular Psoriasis in children

Pustular psoriasis usually occurs as generalized pustular psoriasis (GPP) but can be limited to palms and soles (pustulosis palmaris et plantaris) or to fold areas.[2,3] Four generalized clinical patterns of pustular psoriasis have been described by Baker and Ryan as follows: [4,5]

- Generalized pustular psoriasis of von Zumbusch
- Annular pustular psoriasis
- Exanthematic pustular psoriasis
- Localized pustular psoriasis (not acral and palmoplantar)

These variants are not mutually exclusive and mixed variants (features of both von Zombusch and annular patterns) can occur.[6]

Generalized pustular psoriasis can be divided in two groups which differ in many aspects:[4]

- Patients with a personal history of psoriasis vulgaris (pso + GPP) - corticosteroid withdrawal is the most common precipitating factor, associated with HLA-A1, HLA-B37, and HLA-DR w10.
- Patients without a history of psoriasis vulgaris (pso-GPP) - infections are the most common precipitating factors, associated with earlier age at onset of pustular outbreaks.

Epidemiology

In contrast to nonpustular psoriasis which often presents in childhood and accounts for about 4% of childhood dermatoses, generalized pustular psoriasis is rare in children accounting for 0.6% [4,7,8], posing a diagnostic and therapeutic challenge.[1,9] A history of preceding guttate psoriasis is present in 12% of those who present with GPP in the first decade, in contrast to 85% for adults who present with GPP.[3] Psoriasis vulgaris occurs in more than 59% of patients with generalized pustular psoriasis, and 25% have a positive family history.[10]

Generalized pustular psoriasis in children can occur at any age occurring as early as the first week of life [2] with majority of them being affected between 2 to 10 years of age.[5,10] Generalized pustular psoriasis is slightly more common in boys than in girls with a ratio of 3:2, in contrast to nonpustular psoriasis in childhood and pustular psoriasis in adults.[4,5,12,13]

The von Zumbusch pustular psoriasis is more common in infancy whereas annular forms appear later and mixed patterns also occur. The annular pattern is the most common form observed in children with pustular psoriasis, either alone or mixed with the von Zumbusch pattern.[14] Although localized pustular psoriasis is extremely rare in children, parakeratosis pustulosa is often a manifestation of psoriasis.[11,14]

Etiopathogenesis and precipitating factors

Generalized pustularpsoriasis is a rare inflammatory variant of psoriasis, which presents as systemic inflammatory response syndrome (SIRS).[15]

The aetiology of generalized pustular psoriasis of von Zumbusch type is not fully delineated. Various precipitating factors, most importantly infections (dental and upper respiratory tract infections, group A streptococcal infections, staphylococcal infections), irritating topical treatments like coal tar (Koebner phenomenon) and the withdrawal of oral corticosteroids are well recognized. It is assumed that T cells recognizing bacterial superantigens and/or as yet unidentified autoantigens are involved in the pathogenesis of the disease.[9] The other factors implicated include medications, solar irradiation, pregnancy, emotional stress, vaccination and hypocalcemia. [4,5,7,16,17] The environmental triggers of psoriasis in children are different from those in adults which may explain some of the differences in presentation. Upper respiratory

infections, stress, trauma, and steroid withdrawal are more important precipitating factors in children.[6,12,18] Generalized or annular pustular psoriatic eruptions have occured following Kawasaki disease. [2]

It has been proposed that psoriasis may be a T-cell- mediated autoimmune reaction triggered by a bacterial superantigen and, streptococcal and staphylococcal superantigens have been implicated in the precipitation of pustular psoriasis. While superantigens are clearly able to induce development of skin lesions, a great deal of controversy exists as to whether bacterial superantigens are necessary for disease development. The staphylococcal superantigens include Protein A, cytolytic alpha-toxin, peptidoglycan, and Panton-Valentine Leukocidin (PVL) produced by Methicillin-resistant *Staphylococcus aureus* (MRSA). These superantigens can act as T- cell activators in psoriasis, locally triggering the development of lesions by release of tumor necrosis factor alpha (TNF á) from epidermal keratinocytes. [5,15] The PVL is a cytotoxin representing a stable genetic marker of community acquired MRSA. MRSA is an emerging cause of infection outside of health care settings. Intrafamilial spread of MRSA carrying the gene for PVL has also been reported. Hence it is recommended to perform skin cultures in pediatric patients presenting with new onset pustular psoriasis. Isolation of one infected patient requires investigation of household members and contacts to detect asymptomatic carriage and to treat them.[5]

It has been proposed that the exotoxin produced by group A hemolytic streptococcus acts as a superantigen, and psoriasis persists because of specific T cells that react both with streptococcal M protein and a skin determinant, possibly a variant of keratin. Hence it is suggested to rule out streptococcal infections like pharyngitis, tonsillitis or perianal cellulitis by culture and ASLO titre in patients with pustular psoriasis.[7,17] The recent description of pustules beginning in neonatal period resembling pustular psoriasis clinically and by histopathology, associated with mutations in interleukin 1 (IL1) receptor antagonist, raises the possibility of increased activation of IL1 signalling in pustular psoriasis. This postulation is currently under investigation.[2] It has also been observed that the erythema in pustular psoriasis correlated fairly well with serum IL-6 concentration, suggesting that IL-6 may be a sensitive clinical marker of childhood GPP. [15]

Clinical features

Pustular psoriasis is often the first manifestation of psoriasis in affected infants and children unlike in adult GPP.

Generalized pustular psoriasis was first described in 1910 by von Zumbusch.[7] Many infants with generalised pustular psoriasis have no history of psoriasis vulgaris unlike in adults but 30% have a history of seborrhoeic or napkin dermatitis.[4,7,13,16] GPP is a cyclic process with remissions and exacerbations. The exacerbations exhibit classic phases of the disease, which initially present as sterile pustules formation on a bright erythematous base that coalesce into lakes, subsequently forming the second phase consisting of desquamation. The acute pustulation is painful and usually preceded or accompanied by pyrexia, malaise and anorexia. The acute episode usually resolves within 2–3 days but repeated waves of inflammation and pustulation may follow.[4] Sheets of erythema and pustulation can involve the flexures, genital regions, webs of the fingers and periungual areas. The nails are often thickened or separated by subungual lakes of pus. [2] Mucous membrane lesions are common in the tongue and in the mouth, in the form of geographic tongue, red scaling of lips and superficial ulcerations of tongue and mouth.[2,20] It is considered the most severe form of psoriasis and is associated with systemic associations like hepatitis and arthritis.[20] Most patients with this pattern tend to develop psoriasis vulgaris.[10]

Annular pustular psoriasis, also known as erythema cricine redicivants, Lapiere type psoriasis, erythema annulare centifugum type psoriasis, was first described by Milian and Katchoura in 1933.[6] It is the most common form

observed in children with pustular psoriasis, seen in 60% [14] of cases, either alone (44.5%,) or mixed with von Zumbusch pattern (12.4%),[19] while it is rare in adults. It is characterized by subacute eruption with a pattern of cyclical, gyrate or annular plaques with an erythematous scaly pustular margin, and rapidly evolving pustules that rapidly become desquamative.[6]

Pustulosis palmaris et plantaris is characterized by bilaterally symmetric, chronic pustular eruption on the palms and soles, sometimes associated with psoriasis elsewhere. Deep seated 2-4mm sterile pustules develop within areas of erythema and scaling on the palms and soles. Within few days the pustules resolve, leaving behind a yellow brown scale that is shed within 1-2 weeks. Phases of quiescence and exacerbation are characteristic and exfoliating crusted lesions may occur concurrent with newly developing pustules.[2]

Associations

Pustular psoriasis may be associated with sterile lytic lesions of bone – chronic recurrent multifocal osteomyelitis or SAPHO syndrome, usually affecting the bones of lower limb, pelvis and clavicle.[2]

Complications

Potentially life-threatening complications associated with the disease include, bacterial superinfection, sepsis, dehydration, metabolic, hemodynamic, and thermoregulatory disturbances related to alterations of the epidermal barrier. [2,4,9] Liver abnormalities especially cholestatic jaundice and renal failure have also been described as complications of GPP.[4,]

Course and prognosis

Pustular psoriasis in children tends to follow a more benign but variable course than in adults with spontaneous resolution or development into psoriasis vulgaris.[20] Rarely, it can be serious or associated with chronic morbidity. [5,13,16,19] The course is cyclic, associated with complete clearance of pustular phase and unexplained exacerbations that span decades. Relapses are common and progressively become more severe, often with a poor prognosis. Generalized pustular psoriasis has an explosive course, while localized psoriasis follows a chronic course and portends greater recalcitrance to therapy, and tendency to progress to more widespread disease.[2] Annular pustular psoriasis follows a chronic recurrent course over decades but good prognosis,[6] less severe than those with generalized pustular psoriasis and the chance of spontaneous resolution and long term remission are more in children than in adults. The impact of the disease on the affected child varies according to age, emotional and intellectual development, disease severity, and sites affected.

Differential diagnosis:

The differential diagnosis of childhood GPP includes staphylococcal scalded skin syndrome, impetigo, pustular miliaria, acute generalized exanthematous pustulosis, Reiter's syndrome, pityriasis rubra pilaris, erythrodermic psoriasis, disseminated candidiasis, generalized/ impetiginised atopic dermatitis, seborrheic dermatitis, eosinophilic folliculitis, infantile acropustulosis and subcorneal pustular dermatosis.[2,14,17,20]

In early infancy, accurate diagnosis of pustular skin eruption is especially important, as pustules can be the manifestation of serious infectious diseases or even sepsis. The possible infectious agents implicated in childhood pustular eruptions include varicella zoster virus, herpes simplex virus, meningococcus, staphylococcus, candida, and HIV. Vaccine related diseases, transient neonatal pustular melanosis and deficiency of IL1 receptor antagonist should also be considered.[2,14] DIRA (Deficiency of IL1 receptor antagonist) is an autosomal recessive disorder characterized by psoriasis like pustules during neonatal period associated with joint swelling and pain, sterile multifocal osteomyelitis or periostitis, oral stomatitis, or pyoderma gangrenosum. It is differentiated from psoriasis based on its early onset, poor response to standard therapies of pustular psoriasis, and response to anakinra (1-2mg/kg).[2]

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The differential diagnosis of childhood APP includes tinea corporis, nummular atopic dermatitis, granuloma annulare, urticaria, erythema multiforme, erythema annulare centrifugum, erythema chronicum migrans, subacute lupus erythematosus, ichthyosis linearis circumflexa and annular erythema of infancy.[6]

Treatment

The management of pustular psoriasis in children differs from that in adults in several important aspects including the emphasis on educating the family, dealing with the emotional aspects of the disease and eliminating triggering factors. As childhood pustular psoriasis is a rare disorder, there is a dearth of well designed, comparative and controlled trials on systemic therapy of pustular psoriasis in children, and most of them are empirical based on few case reports with no guidelines or consensus on their use. Hence, children are treated based on the data extrapolated from studies conducted in adults.

Compliance in young children is dependent almost entirely on the parents which is often a reflection of parental motivation. Hence, education of the child and parents is important to encourage a positive approach to the condition and its treatment. Older children and adolescents should be empowered to maintain their own therapeutic routine with parental guidance. The parent and child should be counseled regarding the nature and course of pustular psoriasis with emphasis on the potential triggering factors. They should be made to understand the chronicity of the disease and the tendency for spontaneous remission. The parents should understand the rationale for treatment and should be made clear that the realistic objective is to control rather than to cure the disease. They should be made aware of the various treatment options and their potential side effects and hence the need for continued monitoring. A full informed consent should be obtained especially when initiating potentially toxic agents into the therapeutic regimen and, should be done only by a dermatologist experienced in their use with adequate facilities for monitoring. As far as possible, the treatment regimen should be practical, simple, and not unduly interfering with normal daily activities. It should also be remembered that live vaccines (MMR, OPV) are contraindicated while on cyclosporine and methotrexate therapy. [2] The treatment of generalized pustular psoriasis in childhood is difficult. Owing to the benign course of the disease in most of the cases, it is agreed that the treatment of childhood pustular psoriasis should be conservative and a majority of cases will resolve spontaneously in a few weeks with topical steroids, bland emollients and palliative care. [2,10,17] However, when topical steroids are applied to large areas of the body, hypothalamus–pituitary–adrenal axis suppression and rebound flare on abrupt cessation need to be considered.[14] Compresses in the form of local applications of wet dressings with burrow's solution 1:40 or potassium permanganate 1:5000 (1 crushed tablet 65 mg into 250 ml water) helps to relieve acute flares of the pustular psoriasis.[2] Severe pustular psoriasis, in contrast, is difficult to control and requires hospitalization for supportive management and initiation of a potent systemic treatment regimen with rapid onset of activity to avoid complications. [2,9]

Treatment options in generalized pustular psoriasis in adults include phototherapy, photochemotherapy and systemic agents like retinoids, methotrexate, cyclosporine and biologics. However, clinical experience with the efficacy, systemic toxicity and long-term safety of these agents is extremely limited in children and their potential to cause serious adverse reactions calls for a careful risk/benefit evaluation.[4,9,13] The various therapeutic agents reported to be effective in childhood GPP in literature include oral retinoids (etretinate, acitretin, isotretinoin), methotrexate, cyclosporine, dapsone, steroids , narrow band UVB therapy and biologics (TNF-antagonists – etanercept and infliximab). [1,4,8,13,14,] Most experience has been reported with retinoids which, on the basis of the published data, are considered the first treatment of choice. Methotrexate and cyclosporine appear to be effective in children but more efficacy and safety data are required. [11,16] Annular pustular

psoriasis responds to topical steroids, compresses and if severe, to methotrexate, acitretin and dapsone.[6]

Oral retinoids normalize epidermal differentiation and also have anti-inflammatory activity.[2] They seem to be well tolerated in children than in adults and appear to be safe in children. Hence, they are reported to be the first line of treatment of childhood pustular psoriasis with generally fast response.[4,9] Treatment should be initiated and maintained at dosages at or below 0.5-1mg/kg/day to limit short-and long-term toxicities. When significant improvement occurs, the initial dose should be gradually tapered until a dose of 0.2 mg/kg per day and therapy should be continued for about 2 months after clinical remission. A dose dependent reduction in pustules occurs within few days and the pustules disappear within few weeks, usually between 3 weeks to 4 months.[13] Acitretin is available in 10-mg and 25-mg gelatin capsules and oral administration with milk or fatty foods enhances absorption.[11]

Side-effects are, in general, minor and reversible and include dryness of the skin and mucous membranes and elevation of serum lipids and liver enzymes. Skeletal toxicity in the form of premature epiphyseal closure is a major concern in the treatment of children which is however, very rare when doses do not exceed 1mg/kg/day. [4,9] Hence monitoring with X-rays and growth measurements every 12–18 months has been recommended for children receiving high doses and long term therapy with systemic retinoids. [2,4,11,16] Measurements of base-line lipid profile and liver enzymes are essential. These should be repeated at one-month and then at three-monthly intervals. [2,7,11,16] One of the most important issues is the potential of teratogenicity while using systemic retinoids in the child-bearing population; luckily this side effect can be neglected in childhood. [7] It is imperative that women of child-bearing age are counseled about the absolute requirement to avoid pregnancy during treatment and for 2 years after cessation of the drug in case of acitretin. [16] Hence, it is better to avoid acitretin in sexually active adolescent girls and isotretinoin would be a better alternative as its efficacy is comparable to that of etretinate[13] and contraception is necessary for only 4 weeks after cessation of therapy.[2,13] Despite the wide range of side effects including liver function test and lipid profile abnormality, retinoids are found to be safe even when they are used for periods as long as 6 years.[7] Clinical studies in adults have shown a synergistic therapeutic effect of retinoids in combination with phototherapeutic modalities. Moreover, NB-UVB has been reported to be efficient in the treatment of subcorneal pustular dermatosis, a neutrophilic skin disease related to pustular psoriasis. Hence, acitretin and NB-UVB can be combined for better efficacy. They act in several interlinked ways to inhibit disease activity in pustular psoriasis which includes UVB-induced depletion of Langerhans cells and intraepidermal cells, decreased leucocyte adhesion to the microvasculature, induction of the immunosuppressive cytokine IL 10, retinoid-mediated regulation of keratinocyte growth and differentiation and inhibition of neutrophil function. [9] Based on this synergism, sequential therapy with retinoids and phototherapy would enable optimal control of GPP while minimizing drug toxicity and would be the treatment of choice in children. [21]

Methotrexate is often used as systemic therapeutic agent in adult patients with psoriasis. However, its tolerability and efficacy in childhood psoriasis is still limited. [22] The evidence which is available comes largely from a few studies involving small numbers of patients.[16] Methotrexate is an antimitotic, antichemotactic, anti inflammatory agent usually given at a dose of 0.3mg/kg/week and increased to 0.6mg/kg/week in childhood pustular psoriasis.[2] Methotrexate appears to be a well tolerated drug in children when used in dermatologic doses of 0.2 to 0.4 mg/kg/week and associated with no detectable biochemical or haematological upset. Nausea, gastrointestinal upset, anorexia, fatigue and headache are common adverse effects and bone marrow suppression is an early dose-dependent adverse effect [1,16] These side effects generally respond to temporary cessation of the drug, decreasing the dose or

concurrent folic acid use at a dose of 1-5mg/day on 6 days of the week which decreases nausea, mucosal ulcerations, macrocytic anemia, optimal dosing not determined.[1,2,16] Once clearance is achieved, gradual tapering is done at 2.5mg/month.[2]

The risk of hepatotoxicity is thought to be related to the total cumulative dose. Baseline full blood count, liver enzymes and renal function should be established and then monitored throughout the treatment. Assays are checked weekly initially, then monthly and eventually three monthly. The lack of sensitivity of elevated liver enzymes in detecting early methotrexate hepatotoxicity is a major concern. Some guidelines recommend monitoring by repeated liver biopsies. However, advances in the noninvasive assessment of liver fibrogenesis may eventually reduce the need for liver biopsy. Noninvasive techniques such as dynamic hepatic scintigraphy and serial measurement of serum procollagen III propeptide (P3NP) are of value in monitoring hepatotoxicity. Methotrexate has been detected in liver for upto 116 days after the last dose. Thus intermittent drug free intervals can allow the liver to recover from drug induced toxicities. The fibrotic changes observed in the liver after methotrexate use are potentially reversible. It has been considered that even histological grade 4 liver fibrosis may return towards normal if methotrexate is avoided for 6 to 12 months. The direct markers of hepatic fibrosis in addition to PIIINP include procollagen 1 , type 4 collagen, laminin, hyaluronic acid, tissue metalloproteinases and their inhibitors. There are several important drug interactions with methotrexate (particularly non-steroidal anti-inflammatory agents, antibacterials such as cotrimoxazole, and anticonvulsants such as phenytoin) of which the treating dermatologist should be aware. [16]

Methotrexate is a cheap, easily available, reasonably safe drug having convenient once weekly dose, to be used in severe childhood pustular psoriasis in resource-poor settings. But, there are no guidelines or consensus on the use of methotrexate in childhood psoriasis and is used mostly empirically. However, meticulous use of methotrexate may avert the long- term or serious side effects. [1]

Cyclosporine primarily acts by inhibiting T-cell function and IL 2. It is effective in severe forms of pustular psoriasis when other therapies are ineffective and is often used as a short-term crisis management drug or for the induction of rapid clearance of acute psoriasis flares.[4,11,16] It can be used as single or intermittent short courses as monotherapy and in combination or sequential therapy with other topical and systemic therapies. The rate of improvement depends very much on the dose, which ranges from 3 to 5.0 mg/kg/day.[1,11] After a short period of 3-7 days of cyclosporine treatment, the epidermal mitotic activity of psoriatic skin decreases effectively and lesions usually clear within 2-4 weeks but relapse on stopping.[19] Hence, the dose should be tapered gradually to the lowest dose needed to maintain disease control which usually takes 3-4months.[2] Toxicities of cyclosporine have raised much speculation and prevented its wide usage in childhood GPP, but the benefit/risk ratio is considered acceptable with a short-term, low-dose therapy.[19,21] After remission, a maintenance period is required to prevent focal recurrence of the pustules, but continuation of cyclosporine is not preferred as its prolonged use inevitably results in toxicity and hence a resultant low benefit/risk ratio.[21]

Pharmacokinetics of cyclosporine including intestinal absorption, distribution in body fluids and tissues, metabolism and elimination is age-dependent and hence children need comparatively higher doses of cyclosporine than adults to achieve adequate blood levels with potential risk of dose dependent toxicity. Also, immune reactivity of small children seems to be enhanced necessitating more intensive immunosuppression.[3] However, it has been shown that low dose of cyclosporine are effective in childhood pustular psoriasis.[19] It has also been observed that cyclosporine administration before food produces higher blood concentration without affecting trough levels. Hence, preprandial administration of cyclosporine

would result in a better response.[15]

Dosage adjustments are based on monitoring of clinical response, serum creatinine levels, and blood pressure. Its use is limited by the risk of nephrotoxicity, hypertension and immunosuppression apart from other mucocutaneous side effects. Most of the adverse events are reversible with discontinuation of the medication or a decrease in the dose.[23] No significant adverse events with cyclosporine use in children have been noted on liver function, hematologic values, or development. Nevertheless, cyclosporine must be used with vigilance and adverse events carefully monitored. Although hypertrichosis may be ignored as a mild adverse event, it can be very troubling to parents and older children or adolescents.[23] With the use of cyclosporine in children, concerns about future leukemia, lymphoma, cutaneous carcinoma and oncogenic risks is increased.[2] Thus, in carefully selected and closely monitored patients, cyclosporine can produce relatively rapid clinical response and can be effectively combined with topical and systemic therapies to increase its efficacy and decrease end organ toxicity.[1,11] Biologics are a new treatment option for severe pustular psoriasis, however, very few children have been treated with these agents. Nevertheless, case reports of successful and safe use of infliximab and etanercept in children indicate that biologics may present as promising important treatment option in refractory and severe childhood psoriasis, but their benefit has to outweigh the potential risk of infection, lymphoma and demyelinating disorders.[4,11] There are currently no guidelines for the use of biological agents for psoriasis in pediatric age group and cost is a major limiting factor in most of the developing nations.[11]

Etanercept has been approved in Europe in children above 8 years but not in the USA.[2] It has been used in a double blind randomized trial in psoriasis at a dose of 0.8mg/kg. It may also be a safe and effective alternative for severe palmoplantar pustular psoriasis in children.[24] It has been observed that serious adverse events are rare, and the risk of infection is low because of targeted immunosuppression. Most common adverse events with etanercept include infections (upper respiratory infections, sinusitis), autoimmune complications, and central nervous system disorders such as headaches, mental deterioration, optic neuritis and injection site reactions in the form of redness, pain, and swelling.[23] Etanercept use is also associated with a risk for infections with mycobacteria and salmonella warranting baseline PPD and annual reevaluation.[2] Thus, before initiating and during etanercept therapy, it is important to carefully assess patients for active, chronic, or recurrent infections.[23] Combination therapy of metotrexate and etanercept is associated with increased risk of adverse events, hence etanercept is predominantly used as monotherapy for treatment of dermatologic disorders in children.[23] Although etanercept may provide favorable results, vigilance must be taken when using etanercept in children as the long term risk with its use is unknown. Infliximab also appears promising for the treatment of generalized pustular psoriasis in children, although further experience is warranted and there are reports of pustular psoriasis being precipitated by infliximab. Infliximab can be combined with methotrexate which apart from its synergistic activity, will also take care of the autoantibodies produced by infliximab. Other biologics have not been tried in childhood psoriasis yet.[11]

Dapsone, a relatively safe drug based on a large experience of it being used in many other illnesses has been found to be a moderately effective drug in childhood pustular psoriasis.[8,25]

Psoralen + ultraviolet A (PUVA) therapy is contraindicated in children below 12 years of age [4,10,17] owing to the risk of long-term side-effects, in particular the development of skin cancer,[9] although narrow band UVB (ultraviolet B) has been considered a safer alternative .[4,9,10,17,22] A rare case of acrodermatitis continua of Hallopeau in an infant with an excellent response to the combination of thalidomide and ultraviolet B phototherapy has been reported.[26]

Though systemic steroids have been found to be effective in some cases, most dermatologists are reluctant to use them, as systemic steroids have been found to actually worsen the prognosis and precipitate a rebound flare of GPP when withdrawn or have been ineffective in some cases.[6,10,17]

Conclusion

Childhood pustular psoriasis being a rare disorder, clinical experience with the use of systemic agents is limited. It should be remembered that children are not simply small adults and there is a need for child and parenteral education, compliance, and cooperation as the attitude of the parents and the treating dermatologist can have a major impact on the disability caused by the disease. Thus limited treatment options and compliance are major constraints in the treatment of childhood pustular psoriasis. Hence, more evidence to draw firm conclusions is the need of the hour which should be based on further studies with long-term follow-up periods to define precipitating factors, establish a more definitive cause-and-effect relationship, optimal therapeutic options and safety profiles of different treatment modalities for GPP in childhood. Until a much greater number of children have been treated with these medications, dermatologists must remain vigilant for the potential side effects that have been reported in adults although they may not have yet been reported in children.

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