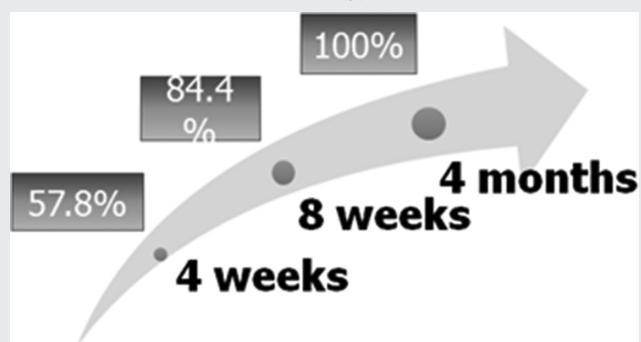
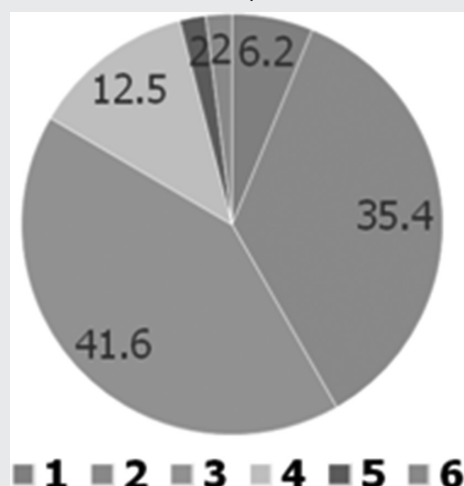


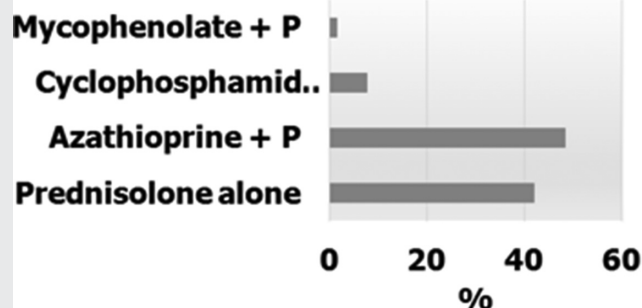
Abstract 365 Table 1 Clinical subtypes of HSPN.



Abstract 365 Table 2 Details of patients with HSPN



Abstract 365 Table 3 Percentage of various clinical subtypes of HSPN.



367 RESTRICTIVE CARDIOMYOPATHY – IS IT THE MOST FREQUENT CARDIAC INVOLVEMENT IN SYSTEMIC SCLERODERMA?

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Background and aims Scleroderma is a rare disease that can involve any internal organ. Systemic scleroderma is usually associated with restrictive cardiomyopathy.

Methods We report the case of a patient diagnosed as having systemic scleroderma since 1975, with cutaneous, pulmonary, digestive involvement, referred to our department for dyspnea on mild exertion, productive cough (mucous sputum), and anterior thoracic pain not related to exertion. All symptoms started about 4 months prior to referral and were very little influenced by corticotherapy and 2 courses of antibiotics administered prior to admission in our department. Clinical examination reveals telangiectasia over the cheek bones, microstoma, spontaneous amputations of distal phalanges fingers 2–4 right hand, basal bilateral pleural friction rub, apical systolic murmur grade 3.

Results Blood examination shows leukocytosis, polyglobulia, dyslipidemia normal ESR and CRP/ECG: sinus rhythm, HR=96/min, QRS axis at +10, LVH, negative T wave (anterior territory). Respiratory functional evaluation shows obstructive dysfunction (reduction by 30% of FEV1). Cardiac

Abstract 365 Figure 1 Timing of renal involvement from onset of HSP

Clinical type	Classification	Laboratory features
1	Isolated hematuria	Urine erythrocyte >5/HPF and proteinuria <150 mg/24hr
2	Minimal proteinuria	150 mg/24hr ≤ proteinuria <25 mg/kg
3	Moderate proteinuria	25 mg/kg ≤ proteinuria <50 mg/kg
4	Massive proteinuria	≥50 mg/kg

Abstract 365 Figure 2 Percentage of various histological grades on renal biopsy.

Parameter	Result
No of HSP	314
No of HSPN	64 (20.4%)
Hypertension	24 (37.5%)
Deranges RFT at admission	5 (7.8%)
Gross hematuria	17 (26.5%)
Relapse	13 (20.3%)
Kidney biopsy	48 (75%)
Death	1
CKD	0

Abstract 365 Figure 3 Treatment in patients with HSPN.

Clinical subtype	Percentage
Massive proteinuria	56.7
Minimal proteinuria	21.7
Moderate proteinuria	15
Isolated hematuria	6.7

ultrasound reveals interventricular septum of 18 mm thickness, posterior wall of left ventricle of 15 mm thickness, with an intraventricular pressure gradient of 60 mmHg at rest.

Conclusions Our patient has the severe, obstructive form of cardiomyopathy. This was the cause of the respiratory symptoms, despite the interstitial pulmonary disease. As a consequence, we changed diltiazem to verapamil, reduced the vasodilators and the corticosteroid doses, which lead to the net improvement of the symptoms. In systemic scleroderma, respiratory symptoms must not be taken “for granted” as being produced by interstitial lung disease., but must be evaluated correctly and completely at each consult.

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IMMUNE THROMBOCYTOPENIA ASSOCIATED WITH LOCALISEDLOCALIZED SCLERODERMA- REPORT OF 4 PAEDIATRICPEDIATRICCASES

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Background and aims Many autoimmune and inflammatory diseases have been found to be associated with morphea. Thrombocytopenia, however, has rarely been reported

Aim-To describe the profile of 4 patients with localised scleroderma and thrombocytopenia

Methods A retrospective case record review of children diagnosed to have localised scleroderma in the paediatric rheumatology clinic of a tertiary care referral institute in North-West India. Children who also had thrombocytopenia (i.e. platelet counts $<150 \times 10^3/L$) were analysed.

Results Twenty two children were diagnosed with localised scleroderma (10 boys and 12 girls, male: female ratio 1:1.2). Thrombocytopenia was identified in 4 children (prevalence-18%). The details of these 4 cases are summarised in Table 1. All 4 cases had en coup de sabre (ECDS) type of linear scleroderma. (Figure 1) Peripheral smear examination revealed normal platelet size. The lowest platelet counts ranged from $8 \times 10^9/L$ to $120 \times 10^9/L$. ANA was positive in 2 patients (case 1 and 2) and case 1 also had positive lupus anticoagulant in her serum. Methotrexate (0.5 mg/kg/week) was used for the treatment of morphea in 3 patients while one child was treated with topical calcipotriol ointment alone. No specific therapy was prescribed for thrombocytopenia, except in case 1 where thrombocytopenia was treated with injection anti D. Platelet counts recovered in 3 patients, while thrombocytopenia is still persisting in one patient.

Conclusions Thrombocytopenia associated with localised scleroderma in children is usually benign, has a probable autoimmune aetiology, requires no specific therapy and is generally curable with the systemic immunosuppressant medications used for the treatment of scleroderma