RHEUMATOLOGY

G163  LONGTERM OUTCOME OF JUVENILE IDIOPATHIC ARTHRITIS: PREDICTIVE FACTORS FOR PAIN

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Aims: To study pain levels and predictive factors for pain in adults with JIA. Methods: Pain was measured by a visual analogue 0-100 scale in 231 adults with JIA. Factors thought to be potential predictors for pain were assessed by forward stepwise linear multiple regression. These included gender, age at disease onset, length of disease, JIA subset, height, growth defects, education, employment, previous depression, clinical joint inflammation (Thompson Kirwan Scale), CRP, ESR, haemoglobin, function (HAQ), mood (HAD Scale), self efficacy (Arthritis Self Efficacy Scale), coping mechanisms (London Coping with Rheumatoid Arthritis Scale) & social contact (Sarason Scale).

Results: There were 63 males and 168 females with a mean age at review of 35.6 years, a mean disease onset at 7.1 years and a mean disease duration of 28.5 years. 37.7% of patients had severe functional limitation (Steinbrocker III or IV), 39.4% had a HAQ score of 1.5 or higher. Only 7% of patients were pain free. In 23.3% of patients, pain was higher than the pain score (0-100). 7% of patients had 90% or better control over their pain (Arthritis Self Efficacy Scale).

The multiple regression analysis identified 5 categories of variables which independently contributed significantly to the variance of pain. In total these variables accounted for 33.5% of the variation in pain.

Cumulative variance R² F p
Function (HAQ) 0.187 48.1 0.001
Control over pain (Self Efficacy) 0.262 37.0 0.000
Joint inflammation (TK index) 0.293 28.0 0.003
Previous depression 0.314 23.5 0.011
Poor coping strategies (London Coping) 0.335 20.7 0.011

Conclusions: Over a third of the variation in pain can be accounted for with 5 independent variables. The variables significantly affecting pain lie within two areas: Disease severity, physical influence on pain and psychological & joint inflammation. Psychological influence on pain - self efficacy, previous depression, coping strategies. This confirms the importance of addressing both physical and psychological factors when attempting to influence a patients’ pain.

G164  LONGTERM OUTCOME OF JUVENILE IDIOPATHIC ARTHRITIS: PREDICTIVE FACTORS FOR MOOD

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Aims: To study mood and predictive factors for mood in adults with JIA. Methods: Mood was measured by a HAD Scale in 231 adults with JIA. Factors thought to be potential predictors for mood were assessed by forward stepwise linear multiple regression. These included gender, age at disease onset, length of disease, number of orthopaedic operations, JIA subset, height, growth defects, education, employment, previous depression, clinical joint inflammation (Thompson Kirwan Scale), CRP, ESR, haemoglobin, pain (pain VAS), function (HAQ), coping mechanisms (London Coping with RA Scale & catastrophising, social contact (Sarason scale), perceived handicap (Disability Repercussion Scale - except emotional handicap subset) & self efficacy (Arthritis Self Efficacy Scale).

Results: There were 63 males and 168 females with a mean age at review of 35.6 years, a mean disease onset at 7.1 years and a mean disease duration of 28.5 years. 31.6% of patients had high anxiety levels, but only 5.2% had high depression levels. However, 23.3% of patients had experienced previous depression (antidepressant use, parasuicide or psychiatric diagnosis). The multiple regression analysis identified 6 categories of variables which independently made a significant contribution (27.6%) to anxiety levels and 5 categories of variables which independently made a significant contribution (46.3%) to depression levels. Depression is mediated by self efficacy symptoms (R²=0.265, F=71.5, p<0.000), perceived social handicap (R²=0.370, F=71.5, p<0.000), age (R²=0.040, F=7.6, p<0.000), social contact satisfaction (R²=0.446, F=41.4, p<0.001) & catastrophising (R²=0.463, F=35.3, p<0.011). Anxiety is mediated by catastrophising (R²=0.144, F=35.0, p<0.000), self efficacy symptoms (R²=0.192, F=24.8, p<0.008), poor coping skills (R²=0.217, F=19.1, p<0.003), inflammation (ESR) (R²=0.240, F=16.3, p<0.002), function (HAQ) (R²=0.260, F=14.4, p<0.018) & social satisfaction (R²=0.274, F=12.8, p=0.048).

Conclusions: Both depression and anxiety are mediated by poor coping skills, reduced self efficacy and low satisfaction of perceived social support. Anxiety which is more prevalent is also mediated by inflammation (ESR) and function (HAQ). Depression is mediated by age and handicap of social activity.

G165  EVALUATION AND TREATMENT OF CHILDHOOD ONSET LOCALISED SCLERODERMA 1988 - 1999

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Aims: To review current practice in the assessment and treatment of children with Localised Scleroderma (LS).

Methods: A retrospective case study from two closely collaborating units. Patients aged <16 years with onset of LS between January 1988 and May 1999 were included. Assessment was by skin score, disease progression, skin texture and joint ROM/contracture, supplemented by information from thermography, clinical photography and MRI scan. Outcome of treatment was determined by improvement in lesion colour, skin texture, joint ROM and lack of progression.

Results: Forty patients were identified, aged 0.9-15.4 years at onset (F:M 3:1). 16 patients had linear scleroderma, 5 scleroderma en coup de sabre, 10 morpha and 9 mixed morpha/linear scleroderma.

In the latter part of the study, 19 patients received combination treatment with methotrexate (Mtx) and intravenous methyl prednisolone (IVMP) and 3 patients received Mtx/oral steroid. The mean dose of Mtx was 9.5mg/m² weekly by mouth (range 2-16mg/m²). The dose of IVMP was 30mg/kg given on 3 consecutive days (7 patients), the remainder received other regimens. Follow-up data is available in 19 of 22 patients. Overall, 12 patients (63%) were “responders”, 3 patients (16%) remained unchanged and 4 (21%) were “non-responders”. Of the 4 non-responders, all have active disease despite the use of multiple other drugs.

Conclusions: The data suggests that the combination of Mtx/IVMP or MTX/oral steroid may be effective in the treatment of LS. Whilst the authors accept the limitations of retrospective analysis, this observation warrants further investigation. A randomised controlled trial is proposed.

G166  POLYARTERITIS NODOSA IN CHILDHOOD

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Introduction: This study describes the clinical, histological, and angiographic features of polyarteritis nodosa (PAN) presenting in childhood.

Methods: Retrospective review of case notes of patients diagnosed with PAN. Only patients who satisfied 3 or more of 10 classification criteria as defined by the American College of Rheumatology (ACR) were included. Angiography was reviewed independently by 2 blinded radiologists.

Results: Between 1971 and 1998, 38 children satisfied 3 or more of 10 ACR classification criteria for PAN. There was a male preponderance of 1:9:1. Mean age was 7.9 years (range 0.3-14.4 years). All had fever and elevation of acute phase reactants. Additional clinical features included rash (61%), renal impairment (24%), hypertension (34%), myalgia (79%), weight loss (79%), testicular pain (20% of males), peripheral neuropathy (13%), cerebral involvement (8%), and sub-acute rhoid hemorrhage (3%). No patient had evidence of hepatitis B infection. 9/12 skin biopsies revealed vasculitis. Renal biopsy was performed in 9 patients and revealed crescentic glomerulonephritis (GN) (4/9), mesangio-proliferative GN (3/9), and focal segmental sclerosis (1/9). Vasculitis was also demonstrated on biopsy of the liver, temporal artery and gut. 35/38 patients had abnormal visceral angiography. A spectrum of angiographic findings was documented and included aneurysms, renal perfusion defects, collateral renal arteries, arterial cut-off, and pruning of the renal arteries. Overall, the mortality for PAN was 8%.

Conclusion: PAN has a wide spectrum of presentation, and is a great imitator of many paediatric conditions. Often the diagnosis remains elusive unless specifically sought, and visceral angiography and tissue biopsy play a key diagnostic role.

G167  CHILDHOOD VASCULITIS IN THE OLD NORTH-WEST REGION

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Background: A new paediatric Vasculitis service started 2 years ago provides care for children in the old North-West region.

Aims: To evaluate the frequency of vasculitis disorders in the old North-West region. To ascertain mode of presentation, time to diagnosis, treatment response and routes of referral in our clinic population.

Methods: 64 children were classified using existing ACR criteria. Children with HSP and Kawasaki’s Disease were excluded.

Results: Systemic Lupus Erythematosus was the most common diagnosis with 11 definite cases and 8 possible evolving cases. 10 cases of systemic onset juvenile arthritis and 10 cases of Juvenile dermatomyositis were included. There were 5 cases of polyarteritis nodosa, 3 sarcoidosis, 2 Wegener’s granulomatosis, 2 ANCA positive vasculitis and 3 with various other defined vasculitis diagnosis. 7 cases of vasculitis were not classifiable and vasculitis was excluded in 3. The majority of patients presented with non-specific symptoms, usually arthritis or skin rash. Some patients had systemic symptoms including fever, weight loss and polyarthritis. Two-thirds of patients had rash. Atrial fibrillation was the commonest cardiovascular abnormality. Aneurysms were seen in 4/11 cases. Visceral involvement was common including liver, intestine and heart.

Conclusion: It is important to maintain a high index of suspicion in the presence of atypical presentations to prevent unnecessary investigations and delays in diagnosis.

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specific symptoms of malaise, high temperature and weight loss. Most patients were referred by general paediatrician and had symptoms for at least 8 weeks. 70% of the patients had at least 2 systems involved. The youngest age of presentation was 14 months of age. Female and male sex ratio (43 Vs 21). One patient died and the group morbidity is high.

Discussion: Children are not classified on the basis of ACR criteria. Concentration of patient's referral allows development of expertise in diagnosis and management. We recommend similar clinics be started in major paediatric centres.

G168 ETHNIC DIFFERENCES IN THE INCIDENCE OF KAWASAKI DISEASE, HENOCH-SCHÖNLEIN PURPURA AND OTHER VASCULITIS IN 664 CHILDREN

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The aim of this study was to determine the incidence and ethnic distribution of Henoch-Schönlein Purpura (HSP), Kawasaki Disease (KD), and rare vasculitides in children resident in one region of the UK.

Methods: Monthly questionnaires were sent prospectively to 239 consultants between 1996-9, with a return rate of 71.1%. Case ascertainment was assessed by review of an additional 393 case notes coded for vasculitis, and by a single questionnaire to 2860 GPs (return rate 59.7%).

Results: 664 new cases of vasculitis in children who fulfilled established diagnostic criteria were collected prospectively from the 1.2 million children resident in the region. Asian and African-Caribbean children had a higher incidence of KD, SLE, and dermatomyositis (JDMs) than Caucasian children.

| Table: Incidence of vasculitis per 100,000/year (ethnic distribution (%)) |
|------------------|----------------|----------------|----------------|
| Ethnic Distribution | KD  | HSP  | SLE  | JDMs |
| Total             | 2.97 | 21.1 | 0.8  | 0.4  |
| Asian             | 10%  | 5.9  | 25.3 | 12.8 | 386  | (n = 508) |
| Black             | 8%   | 1.7  | 2.5  | 1.7  | 169  | (n = 28)  |
| Caucasian         | 82%  | 1.1  | 24.4 | 0.4  | 43   | (n = 15)  |

The incidence of KD rose to 5.6/100,000/year in children under 5 years, with the same ethnic distribution. Five children had coronary artery involvement, of whom 2 were Asian. The incidence of HSP (determined over 2 years) was higher than previously reported. 3 cases, all Asian, of polyarteritis nodosa (PAN), and single cases of Wegener’s granulomatosis (WG) (Caucasian), Behcet’s (Caucasian) and microscopic polyangiitis (Asian) were identified. The rare vasculitides had a high complication rate with 2 deaths (both PAN), renal failure (28% of SLE, and the case of WG), and cerebrovascular occlusion in 25% of SLE.

Conclusion: Our study has recognised a high incidence of vasculitis in the Asian and African-Caribbean populations, with a high morbidity and mortality associated with the rare vasculitides.

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G169 THE PHYSIOTHERAPY MANAGEMENT OF REFLEX SYMPATHETIC DYSTROPHY

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Methods: Records of all children with RSD attending the physiotherapy department of this hospital were reviewed. Children had been medically assessed and investigations completed prior to referral to the department. Management consisted of an explanation of RSD and its link with stress; an individualised exercise programme and time spent with each family exploring potential stress factors. Records were reviewed to assess response to treatment, time to response and relapse rate.

Results: 53 children were treated in the period of study. Male to female ratio was 7:46. Age range was 8-16 with 10 aged 8-10 years; 25 aged 11-13 years and 18 aged 14-16 years. All responded to treatment, 9 either responded partially or subsequently relapsed. No other treatments such as guanethidine block.

Conclusions: RSD is common in children and responds well to appropriate physiotherapy management. This includes an exploration of underlying triggers and involvement of the Child Psychiatrists as necessary.

G170 INTRA-ARTICULAR STEROIDS AND EARLY METOTREXATE GIVE A SUSTAINED IMPROVEMENT IN JUVENILE IDIOPATHIC ARTHRITIS (JIA)

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Introduction: Intra-articular triamcinolone hexacetonide (IATH) and intra-venous methyprednisolone have both been used to control polyarthritis in JIA. We reviewed our recent experience of both these approaches to try to ascertain their effectiveness.

Methods: Over an 18 month period 119 children with JIA had 168 joints injected and 11 were given IVMP. The mean dose of triamcinolone hexacetoni- lide used was 0.07 mg/kg (0.04-0.21) for large joints and 5 mg/joint (1-8) for small joints of the hand. 3 daily pulses of 30 mg/kg (max 1g) of mpiethpredni- solone were used. 50% were taking MTX and a further 29% were commenced on MTX at the time of IATH. Mean dose of MTX was 0.51 mg/kg week (64% subcut, 36% oral).

A fall in ESR and AUC were seen in children who received IATH although a mean of only 31/3 active joints was injected. The response in the IVMP group was comparable. Those taking MTX had a sustained response over 9 months, while the AUC in children not receiving MTX started to rise by 9 months.

Conclusion: IATH and IVMP when used in conjunction with MTX both produce a sustained response. We propose a blinded, randomised, prospective trial to look for any advantage between the IATH and IVMP.

G171 HOME ADMINISTRATION OF SUBCUTANEOUS METOTREXATE IN JUVENILE IDIOPATHIC ARTHRITIS

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Methotrexate is an effective treatment for juvenile idiopathic arthritis. Parenteral administration is used where there is an intolerance of, or poor response to, oral therapy or a need to use larger doses. The subcutaneous route is preferred to the intramuscular route giving equivalent bioavailability but less pain. Home administration of methotrexate by the family is possible for many children and improves quality of life, removing the need for weekly hos- pital or clinic visits.

We describe the development of a protocol that addresses many of the health and safety concerns surrounding the home administration of subcuta- neous methotrexate. This includes a programme of training for the parents regarding health and safety issues in addition to the practicalities of adminis- tering the injections.

Since the completion of the protocol, 11 parents have been trained to adminis- ter methotrexate at home. Training has been uneventful. All parents have been satisfied with the training and confident in administering the drug at home. No health and safety concerns have been raised.

Subcutaneous methotrexate can be safely administered at home provided families receive appropriate training, and measures are in place to address health and safety concerns.

G172 METOTREXATE USE IN PAEDIATRIC RHEUMATIC DISEASE: A 10 YEAR EXPERIENCE

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Aims: This study examined trends in Methotrexate (MTx) use in a tertiary centre for paediatric rheumatology. Knowledge of VZ immunity in this patient group was reviewed.

Methods: A prospective and retrospective case record study identified patients receiving MTx between March 1989 and November 1998. Patient demographics, diagnosis and details of methotrexate prescription, route of administration and monitoring were recorded. Evidence of VZ exposure was sought from clinical history and microbiological records.

Results: 118 patients were identified, of whom 106 were receiving MTx at the time of the study (age range 2.2 - 25.6 years). Sex distribution: F:M 65:35. Potential carrier (JIA Rh fac –) and positive Systemic Onset (JIA) represented 34% and 28.3% respectively. The number of patients starting MTx increased exponentially over the study period (1989 n=1; 1998 n=24). The time between diagnosis and starting MTx fell from 2.2 to 0.36 years between 1989 and 1998. The oral route of administration was most common, however increasing subcutaneous (SC) use was documented. In 1998, 46 (43%) patients received SC MTx, 10 self administered, 20 were administered by a relative and 16 by a practice/practice nurse. Blood monitoring was undertaken by the tertiary centre.
in 24.5% patients, 20.7% local hospital, 54.8% GP. VZ status was recorded in (51.9%) patients.

**Discussion:** Mtx is increasingly used in paediatric rheumatic disease. Trends towards earlier and more aggressive intervention are noted. It is recommended that VZ IgG titres are determined prior to initiation of Mtx therapy.

**G173**

**THALIDOMIDE USE IN CHILDHOOD BEHÇET’S SYNDROME**

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**Objective:** To study the clinical spectrum of Behçet’s disease (BD) in childhood, and to report our experience of using thalidomide.

**Method:** Ten children, who were diagnosed as BD, were studied retrospectively. Nine fulfilled the international criteria of complete BD and one was partial BD.

**Results:** Mean ± SD at first presentation was 5.4 ± 3.8, at diagnosis 10.1 ± 3.8 and follow-up period for 3.8 ± 1.7. Oral ulcers were present in all patients (100%), Genital ulcers in 6 (60%), peri-anal ulcers in 3 (30%), skin manifestations in 9 (90%), intra-cranial hypertension in 2 (20%), mild gastro-intestinal symptoms in 5 (50%), joint symptoms in 6 (60%), ocular lesions in 5 (50%), but only one child had anterior and posterior uveitis. Colchicine was used in 5 children with good response and thalidomide in 5 children who did not respond to other immuno-suppression agents at mean age 12.2 ± 3.4 years. Thalidomide (1mg/kg/week to 1.1 mg/kg/day) resulted in complete remission in 3 children and less frequent milder oral ulcers in 2 children. Neupathy developed in 2 children, one at a dose of 0.6 mg/kg/day and was reversible, the other was irreversible at a dose 1 mg/kg /day.

**Conclusion:** BD in children is similar to the disease in adults. Thalidomide provided a useful therapeutic option in severe oral and genital ulceration, which was unresponsive to other therapies. Awareness of the danger of axon neuropathy and teratogenesis at all times during thalidomide therapy is crucial. A low dose of 1-2mg/kg/week is probably as effective as higher doses.