ISOLATED CENTRAL NERVOUS SYSTEM (CNS) VASCULITIS IN CHILDHOOD - A REPORT OF FIVE CASES WITH LONG TERM FOLLOW-UP. George L. Allen, Audrey M. Nelson, Manuel R. Gomez. Mayo Clinic, Rochester, MN 55901, USA.

Vasculitis isolated to the CNS without apparent systemic involvement is unusual. We report five cases of this entity with long term follow-up. Each case had the characteristic lesions of CNS vasculitis demonstrated on cerebral angiography. The age of onset was from 3-15 years. There were three females and two males. Each had a hemiparesis which came on suddenly or developed over a two to three day period as the chief manifestation. Prodromal symptoms included headache in two cases, slight dizziness in one case and undue fatigue in another. Each patient had demonstrable neurologic deficits on physical examination and none had any of the usual manifestations of systemic vasculitis such as myalgias, peripheral neuropathy, arthralgias or other organ involvement. Basic laboratory tests were normal including CBC, ESR, FANA, anti-n-DNA, LE prep, serum protein electrophoretic pattern, and CSF studies. EEG and head CT when done demonstrated abnormalities expected from the physical exam and cerebral angiography.

These cases have been followed 8-18 years (mean 10.8 years) without evidence of further vasculitis and in each case progressive recovery was observed. This recovery was virtually complete in two cases; there was mild residual weakness in one case, and a moderate residual hemiparesis in two cases. Three of the five cases were treated with steroids. None received immunosuppressive agents.

Long term follow-up would indicate that isolated CNS vasculitis in childhood may carry a more favorable prognosis than previously thought.

A LONG-ACTING ANTIINFLAMMATORY DRUG, OXAPROZIN, IN THE TREATMENT OF JUVENILE RHEUMATOID ARTHRITIS (JRA). J.C. Bass, Ohio State University, Columbus, OH 43205; B.H. Athreya, University of Pennsylvania, E.J. Brewer, Baylor College of Medicine; D.P. Goldsmith, Temple University, School of Medicine; J.R. Hollister, National Jewish Hospital; D.W. Kredich, Duke University Medical Center; N.E. Brandstrup, J.J. Miller, Stanford University School of Medicine; L.M. Pachman, Northwestern University. The Pediatric Rheumatology Collaborative Study Group, U.S.A.

Fifty-nine patients with JRA were enrolled in a three month open-label, multi-center study of oxaprozin. A once daily dosage schedule based on body weight began at 10 mg/kg/day and was increased to a maximum of 20 mg/kg/day (range 10-24). Thirty patients continued the maximum dosage into a nine month extension period. Global assessment scores of the investigators, parents and patients showed moderate to marked improvement in more than 60% of patients. At least 25% improvement from baseline values occurred in more than 30% of these patients. Significant mean changes were apparent in total score for joint swelling, active joints, grip strength, travel time and duration of morning stiffness. Results were maintained during the extension period. At least one drug related adverse effect was reported by 47% of patients during both parts of the study with gastrointestinal disorders being the most common. Eight patients discontinued treatment because of drug-related problems. A vesicular eruption on exposed skin areas that increased with duration of treatment was seen in 10 patients, significant decreases in hemoglobin in 5, and increases in transaminase levels in 2. Oxaprozin appears to be more phototoxic but otherwise similar to other non-steroidal antiinflammatory drugs in children with JRA.

NAPROXEN (NAP) AND ACETYLSALICYLIC ACID (ASA) IN THE TREATMENT OF JUVENILE RHEUMATOID ARTHRITIS (JRA). DOUBLE BLIND PARALLEL 24 WEEK TRIAL. Tore K. Kvien, Hans M. Høyeraal, Berit Sandstad, and Erik Kåss. Oslo Sanitetsforening Rheumatism Hospital, Oslo, Norway.

Eighty patients with JRA were randomized, 40 in each group. Female: male 3:1. Median age was 12.4 yrs, range 3.2-15.7. Pauci:polyarticular type, onset 2.3:1, course 1.8:1. NAP was given b.i.d. 10mg/kg/day and microencapsulated ASA t.i.d. 75mg/kg/day. All antirheumatic therapies were stopped for defined periods of time prior to randomization. Concomitant drug and surgical therapy were not allowed. Changes in disease activity were mainly assessed at 12 and 24 weeks. Median values are given. The number of patients is given in brackets.

Disease activity	Baseline = 0		12 weeks - 0		24 weeks - 0	
	NAP(40)	ASA(40)	NAP(36)	ASA(23)	NAP(25)	ASA(16)
Subjective (x)	4	3	0	0	0	0
Physician (x)	8	8	-2	-1	-2	-2
Active joints (y)	55	50	-12	-2	-21	-11
Swollen joints (y)	50	50	-13	-2	-14	-11
Grip strength (z)	87	70	6	3	15	8

x) Global assessment by graphic rating scale (1-20, 20=maximal activity) y) Index, grading joints by severity and size. z) in mm Hg.

Improvement of disease activity was seen in 23 NAP treated patients and in 16 with ASA, lack of response in 12 NAP and 2 ASA. Withdrawals due to adverse reactions were more frequent in ASA treated patients (20) than in NAP (5), gastrointestinal in 7/2, CNS 12/2, skin 3/2, liver 8/0.

Both drug regimens seemed therapeutically effective, but NAP resulted in fewer adverse reactions than ASA.

JRA-ADJUSTMENT TO ILLNESS. Raquel V. Hicks, Solange Cook, Willow Morton, Gayle Kutaka. University of Hawaii, Honolulu, HI USA.

Psychological adjustment to JRA was studied in 24 children (5-18 yrs) living at home by a) psychological battery, b) structural interview, c) standardized behavioral observations, d) school report. A marked increase in behavior disorders (B.D.) was present at onset of JRA and at assessment compared with pre-illness:

	Pre-Illness		Onset JRA		Current	
	N	%	N	%	N	%
No Disorder	16	67	9	38	10	42
Disorder	4	16	13	54	14	58
No Information	4	16	2	8	0	0

Diagnoses were dysthymic behavioral disorder (B.D.) 29%, oppositional B.D. 21%, conduct B.D. 21%, and one each educable MR, phobia, schizoid B.D., malingering and adjustment reaction. 20/23 siblings and 21/30 parents were free of B.D. Bright average and superior I.Q. prevailed but 45% were underachieving, 41% expected achievement, 12% overachieved relative to potential. B.D. was significantly and directly related to parent adjustment and overprotectiveness. The frequency of B.D. tended to be directly related to age and severity of impairment. Children perceived their illness as active aggression requiring energy to counteract. Excessive anxiety, poor anger control, self-blame, dependency, constricted personality were prevalent.

Areas identified in this study provide direction for developing preventive therapeutic programs which should be started at the onset of JRA. Adolescents, the severely disabled and those with poorly adjusted, overprotective parents are at highest risk for B.D.