1033 DECREASED CMI, ABNORMAL MACROPHAGES AND INTERFERON FUNCTION IN ANHYDRITIC ECTODERMAL DYSPLASIA. James F. Jones, Vincent A. Fulginiti. Univ. of Ariz., University Hospital, Dept. of Pediatrics, Tucson.

Ectodermal dysplasia syndromes are associated with varying degrees of cellular immune defects. Two male siblings with anhydritic ectodermal dysplasia (AED) succumbed to generalized atypical tuberculosis infections. The second child had recurrent bacterial infections as well and is the subject of this report. At age 10 years skin anergy to multiple antigens was present when M. intracellulare was isolated from bone marrow, spleen, liver and multiple lymph nodes. Evaluation of immune function included T and B cell enumeration, mitogen and antigen stimulation, MIF production, serum immunoglobulin and specific antibody production, PMN and macrophage phagocytosis, killing and chemotaxis, and lymphocyte interferon production. General anergy and depressed PMN and macrophage chemotaxis varied indirectly with positive responses to antimicrobial therapy. Persistent abnormal responses included no skin test or in vitro lymphocyte response to M. tb or M. intracellulare, abnormal macrophage morphology, poor granulomata formation, and absent interferon (IF) production. Patient lymphocytes failed to produce IF even after contact with normal macrophages. Although normal lymphoid tissue was present at diagnosis, no lymphoid tissue was present at post mortem. Afferent and efferent defects in immune function appear to be responsible for this fatal illness. Further study of this axis in recovery from tuberculosis and in AED may elucidate the primary or secondary nature of such defects.

1034 COMBINED CMV-EBV INFECTION TREATED WITH ORAL TRANSFER FACTOR (OTF). James F. Jones, Linda Minnich, Wayburn Jeter, and Vincent A. Fulginiti, Univ. of Arizona, Departments of Pediatrics, Pathology and Microbiology, Tucson.

A 4-year old male had recurring episodes of arthritis, skin rash, abdominal pain, vomiting and atypical lymphocytosis for 1½ years following an acute illness in Jan. 1978 that included pharyngitis. No etiology was found during the first 2 months of illness; further serological testing identified the probable cause as EBV. Fluctuating CMV titers suggested a dual infection.

TITERS:	3/15/78	4/6/78	4/20/78	5/6/78	4/15/80
EB VCA	128	256	128	256	20
EBNA	2	8	16	4	2
CMV	128	32	1024	512	320

Viruria was first detected in April 1978 and persisted for 15 months with a peak titer of 10^{10} INFU/ml in March 1979. CMV was present in a buffy coat culture during an episode in Nov. 1978. Although CMV titers were persistently increased, in vitro lymphocyte responses to CMV remained negative (SI <1.0; control >3.0); responses to mitogen were normal. A 7-month trial of levamisole was ineffective. Bovine OTF from an animal skin test positive to infectious rhinotracheitis virus (cross reacts with human CMV) was administered every three weeks for 6 months. Two months after starting OTF, symptoms and viruria ceased and CMV SI was 4.0. He has remained symptom-free with a continued positive SI for 1 year without further therapy. Recovery from this chronic CMV infection was temporally related to OTF administration. Further evaluation of treatment with OTF for chronic virus infection accompanied by abnormal cellular immunity is suggested.

1035 FACTORS INFLUENCING DEVELOPMENT OF GROUP A STREPTOCOCCAL TYPE-SPECIFIC ANTIBODY. Edward L. Kaplan, Aurea Flores, Dwight Johnson, Lewis W. Wannamaker. University of Minnesota, Dept. of Pediatrics, Minneapolis.

Because factors influencing the immune response to streptococcal M protein following upper respiratory tract (URT) and skin infections are poorly understood, we evaluated epidemiologic data from 4 groups of patients (408 serially collected sera from 119 children; $\bar{m}$ age = 6.5 yrs.) followed carefully for 2 yrs. The date and site of first acquisition of 4 serotypes (6,31,49,52), and the type-specific serum bactericidal activity (SBA) from pre- and post-acquisition sera were determined. The effects of patient age, serotype, site of isolation and number of culture-positive visits for each M type on the development of SBA were studied.

M-6 was isolated from only the URT; while M-31, 49, and 52 were isolated from both skin and URT. Significant SBA developed in 27 of 41 (66%) patients with M-6; 22 of 41 (54%) with M-31; 13 of 31 (42%) with M-49; and 4 of 6 (67%) with M-52. The magnitude of the SBA response was generally greater for those with M-6. Of those with M-31 or M-49, no difference in SBA was noted between those with recovery only from the URT and those with recovery from both the URT and skin lesions. The development of SBA was related to number of isolations, although different for those acquiring each type. The increase in SBA was age-related in those with M-6, but not in those with M-31 or M-49. These data define several complex variables which may influence development of SBA following group A streptococcal infections.

1036 THE EFFECT(s) OF FACTOR(s) GENERATED BY INVADING PATHOGENS ON CHEMOTAXIS (CX) OF HOST NEUTROPHILS (PMNS). Abdul J. Khan, Mathew Varghese, Kusum Kumar, Urmila Patel and Hugh E. Evans, Dept. of Ped., Jewish Hospital and Medical Center, Brooklyn, New York.

The effect of bacterial infection on PMN CX has not been studied in detail. Whether infection causes impairment or enhancement is controversial. The response during infection, to chemotactic factors (CF) generated by the pathogen is unknown. 14 patients (mean age 4.5 yrs) and 14 age matched controls were studied. 4 had septicemia; 1 meningitis; 5 UTI, 6 Enteritis and 1 otitis media. The organisms isolated (Pneumococcus, Shigella, Pseudomonas, E. coli, Klebsiella and Salmonella) were grown in broth. CF for each organism (OCF) was obtained. 12 patients were studied before and 2 during antibiotic therapy. CX was performed utilizing Boyden's technique. PMNS placed on 3μ micropore filter were tested against OCF, endotoxin activated serum (EAS) and an standard E. coli CF (ECF) to determine chemotactic index (CI) and against Hank's solution random migration (RM). The mean (± 1SD) CI and RM are presented (table). CI obtained with

	OCF	EAS	ECF	RM
Patients	28(9)	31(10)	31(11)	22(13)
Control	51(11)	57(13)	58(14)	18(6)
P Value	< 0.002	< 0.0005	< 0.005	> 0.1

the 3 CFs were decreased and RM unchanged compared with controls. These findings suggest that the invading organism contributes to the host's vulunerability and may impair recovery by decreasing CX.

1037 ADHERENCE OF HAEMOPHILUS INFLUENZAE b TO EPITHELIAL CELLS OF CHILDREN. Richard M. Lampe, Sheldon L. Kaplan, Edward O. Mason, Jr., Carol L. Umstead, Martha D. Yow, and Ralph D. Feigin. Baylor College of Medicine, Texas Children's Hospital, Department of Pediatrics, Houston, Texas.

Bacterial adherence to epithelial surfaces may play a role in the colonization and pathogenesis of bacterial infections. Using fluorescent antibody and radiolabeled methods, we have demonstrated that H. influenzae non-typeable (HiNT) adhere more readily to buccal epithelial cells (BEC) than do H. influenzae b (Hib). The adherence or non-adherence of Hib strains was not influenced by BEC source (different adult donors) or site (pharyngeal, nasal or buccal) of the same adult donor, antibiotics or heat-treatment of BEC. One percent mannose, ribose and dextrose inhibited the adherence of the one adherent Hib strain (throat).

BEC from 10 asymptomatic children, 8 with confirmed systemic Hib disease, and 10 with symptomatic respiratory infections (URI) were studied using one adherent and three non-adherent Hib strains (2-CSF, 1 ear). Nasal cultures for viruses were positive in 1/10 with URI. The throat strain adhered equally to BEC from all three groups and the non-adherent Hib strains did not adhere to the BEC from 10 control children, the 10 children with symptomatic URI and 7/8 children with confirmed Hib diseases. One child studied twice while asymptomatic and symptomatic (URI) showed no change in Hib adherence.

This diminished adherence of Hib strains to epithelial cells may be relevant to the invasiveness of Hib and to the pathogenesis of Hib infection.

1038 PEDIATRIC NOCARDIOSIS; Barbara J. Law, Melvin I. Marks. Univ. of Oklahoma Health Sciences Center, Dept. of Pediatrics, Oklahoma City, Oklahoma.

Nocardial infections are considered rare in children, occurring mainly as pulmonary disease in immuno-compromised hosts(CH), with N. asteroides the predominant species. A review of our experience over a five year period('75-'80) suggests a different clinical spectrum. A summary of the clinical data is shown below:

Isolate	n	ages	sex (M/F)	CH	Primary site lung/CNS/skin	Deaths
N. asteroides	5	2mo-18yr	1/4	4/5*	3 / 1 / 1	3
N. brasiliensis	5	2-5yr	5/0	0/5	0 / 0 / 5	0

* 1 chronic granulomatous disease, 1 immunodeficiency, 1 lymphoma, 1 hydrocephalus with ventriculoperitoneal shunt.

Of special interest was the frequency of N. brasiliensis infection, formerly considered very rare in the U.S. and usually manifest as mycetoma. In contrast, all 5 patients were previously healthy children who developed acute subcutaneous abscess(2 submandibular, 1 scalp, 1 axilla, 1 hand & elbow); 2 had a primary pustular lesion with regional suppurative adenitis; 3 had a history of trauma. Gram stain of purulent material showed gram-positive branching filaments in 2 of 3. Therapy(drainage and/or antibiotics) was successful in all 5. We conclude that nocardiosis, particularly that due to N. brasiliensis, is more common in normal children than previously reported and should be considered in the differential diagnosis of subcutaneous abscess, especially if there is a history of trauma.