



Hyper IgM Registry Data Collection Form

Patient Initials: ___/___/___

PLEASE NOTE: NEMO HAS IT'S OWN FORM

Patient Identification:

Patient Name (first, middle, last) _____

Patient's USIDNET Registry Number assigned after online enrollment _____

Date of Birth ___/___/___ (mm/dd/yyyy) or Year of Birth _____

Gender: male [], female []

Home Address:

Address: _____

State: _____
Zip Code: _____
Phone: _____
Email: _____

State or Province of birth: _____
Country of birth: _____

Date of this Record Completion (mm/dd/yyyy): ___/___/___

Date of Visit (mm/dd/yyyy): ___/___/___

Is this the initial registration of this patient[] or follow-up?[]

Submitting Physician Information:

Name: _____
Address: _____

State: _____
Zip Code: _____
Phone: _____
Email: _____
Fax: _____

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Spectrum of Disease

HIGM-1/HIGM-3: Male patients with XHIM (HIGM-1, CD40L defect) or male/female patients with HIGM-3 (CD40 defect) have recurrent bacterial and opportunistic infections starting in the first year of life. Pneumocystis carinii pneumonia is a common presenting infection. Other patients may have chronic, profuse diarrhea requiring parenteral nutrition. Over 50% of patients have chronic or intermittent neutropenia, often associated with oral ulcers. Cryptosporidium infection may lead to severe bile duct disease and hepatic cancer. Serum concentration of IgG is usually less than 200 mg/dl; IgM may be low, normal or elevated. Atypical cases may present with recurrent infections, anemia or hepatitis in the second or third decade of life.

AID/UNG defect: Male/female patients with AID (HIGM-2) or UNG defects have recurrent bacterial infections without opportunistic infections. The majority of patients have lymphoid hyperplasia and many patients may develop an autoimmune disease such as diabetes mellitus, polyarthritis, autoimmune hepatitis, chronic uveitis, or Crohn's disease.

Diagnostic Criteria (HIGM)

Definitive HIM []

Serum IgG concentration at least 2 SD below normal for age [] and **one of the following**:

Mutation in any of the following genes

CD40L (XHIM, HIGM-1)	[]	AID (AR-HIM,)	[]
CD40 (AR-HIM, HIGM-3)	[]	UNG (AR-HIM	[]

Maternal cousins, uncles, or nephews with confirmed diagnosis of XHIM []

Siblings with confirmed diagnosis of AID [], CD40 [], UNG []

Probable/Possible HIM []

Serum IgG concentration at least 2 SD below the normal for age with normal T and B cell numbers []

AND (select all that apply):

Serum IgM concentration at least 2 SD above normal for age []

Normal T cell proliferation to mitogens []

No antigen specific IgG antibody []

Absent switched memory B cells []

Recurrent bacterial infections in the first 5 years of life []

Pneumocystis carinii infection in the first year of life []

Neutropenia []

Cryptosporidium-related diarrhea []

Sclerosing cholangitis []

Parvovirus induced aplastic anemia []

Lymphoid hyperplasia (AID/ HIGM-2 or UNG defect) []

Other _____

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Additional Clinical Features of Special Note List all complications in the comment field at the end of this section *.

	Not Seen	Observed	Prominent in this Patient	Unknown
Lymphoid hyperplasia				
Neutropenia				
Sclerosing Cholangitis				
Arthritis, chronic RA/polyarthritis				
Arthralgia				
Vasculitis				
- Skin				
- Coronary				
- Renal				
- Cerebral				
- Pulmonary				
- GI				
- Other (specify)				
Nephritis				
Chronic uveitis				
Enterovirus encephalitis				
Unexplained CNS deterioration				
Osteoporosis				
Other (explain)				
Comments:				

Autoantibodies

	Yes	No	Unknown	Not Tested
ANA positive				
Anti-neutrophil antibody				
Anti-platelet antibody				
Coomb's positive (direct)				
Other auto-antibodies (specify) _____				

Other treatments used List all complications at the end of this section *.

	Yes	No	Unknown	Age begun		OR Date begun (mm/dd/yyyy)	Duration	Still needed?	
				Mos.	Yrs.			Yes	No
Chronic oxygen									
Parenteral nutrition									
Other:									

Other? Please explain _____

***** Complications to any surgery/treatment listed above (list and explain):**
