

CVID International Splenectomy Survey Patient Survey Form

Patient ID

Center	1. initial- 1. and last surname	Year of birth	Sex <input type="radio"/> male <input type="radio"/> female
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Section 1. Diagnosis of CVID

a) Year of CVID diagnosis Unk

b) Clinical manifestations

	Yes	No	Unk		Yes	No	Unk
Splenomegaly	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	AI cytopenia AIN, AIHA, AITP (please circle)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Lymphadenopathy	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	ILD	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Granulomatous disease	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	Liver disease	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
GI disease (please specify)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	Other (please specify)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

c) Immunoglobulin levels pre- Immunoglobulin replacement therapy Unk

IgG g/l	IgA g/l
IgM g/l	IgE g/l

d) Immunoglobulin replacement therapy before splenectomy Yes No Unk

Route sc iv Unk

Average dosage of monthly replacement

IgG level under standard replacement g/l

mg / kg body weight

e) Lymphocyte phenotype before splenectomy Unk

Lymphocyte count / μ l

Date Month Year

CD3+ / μ l	CD4+ / μ l	CD8+ / μ l
CD19/20+ 	CD3-, 16/56+ 	

f) CVID classification

Freiburg <input type="radio"/> Group Ia	Paris <input type="radio"/> MB 0	Euroclass <input type="radio"/> B -
<input type="radio"/> Group Ib	<input type="radio"/> MB 1	<input type="radio"/> B + <input type="radio"/> smB- <input type="radio"/> Tr norm <input type="radio"/> 21 norm
<input type="radio"/> Group II	<input type="radio"/> MB 2	<input type="radio"/> smB+ <input type="radio"/> Tr high <input type="radio"/> 21 ^{low}
<input type="radio"/> Unk	<input type="radio"/> Unk	<input type="radio"/> Unk

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g) T cell and B cell profile before splenectomy

T cell profile

Date
Day Month Year

		% of lymphocytes	absolute (/ μ l)	unknown
CD3 total	Total T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of T cells		
CD4	CD4 T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD8	CD8 T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD3+ TCR α/β + CD4/8-	Double negative T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of CD4 T cells		
CD4 CD45RO	CD4 memory T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of CD4 CD45RO T cells		
CD4 CD45RO CXCR5+	Circulating TFH like CD4 T cells	<input style="width: 100px; height: 20px;" type="text"/>		<input type="radio"/>
		% of CD4 T cells		
CD4 CD45RA CD31+	CD4+ Recent thymic emigrants	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of CD8 T cells		
CD8 CD27- CD28-	Late CD8 effector T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of CD8 T cells		
CD8 CD27+ CD28-	CD8 effector T cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>

B cell profile

Date
Day Month Year

		% of lymphocytes	absolute (/ μ l)	unknown
CD19 Total	Total B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
		% of B cells		
CD27- IgM+ IgD+	Naive B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD27+ IgM+ IgD+	IgM mem / Marg. zone B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD27+ IgM- IgD-	Switched mem, post GC B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD38 ++ IgM ++	Transitional B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD38 +++ IgM -	Plasmablasts	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>
CD21 ^{low} CD38 ^{low}	CD21 ^{low} B cells	<input style="width: 100px; height: 20px;" type="text"/>	<input style="width: 100px; height: 20px;" type="text"/>	<input type="radio"/>

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Section 2. Splenectomy

a) Year of diagnosis of splenomegaly Unk b) Year of splenectomy

c) Why was splenectomy performed?

d) What other treatment was tried prior to splenectomy?

e) What was the outcome of that treatment?

f) Spleen size Spleen weight Unk Ultrasound measurement Unk
 g diameter cm x length cm

g) Spleen histology

h) Any surgical complications?

i) Did the patient receive prophylactic antibiotics post splenectomy? Yes No Unk

If Yes, for how many month?

Section 3. Status of patient since splenectomy

a) Follow up after Splenectomy months

b) Effect of splenectomy on problem identified in 2 c)

c) Patient deceased? Yes No Unk

Date
Day Month Year

Cause of death _____

d) Any problems with severe infections?

Please give details including dates, infection type and organisms.

e) Immunoglobulin replacement therapy after splenectomy Yes No Unk

Route sc iv Unk

Average dosage of monthly replacement

IgG level under standard replacement g/l

mg / kg body weight

f) CVID classification

Freiburg Group Ia Paris MB 0 Euroclass B -
 Group Ib MB 1 B + smB- Tr norm 21 norm
 Group II MB 2 smB+ Tr high 21^{low}
 Unk Unk Unk

g) Overall evaluation of outcome after splenectomy

excellent good mixed poor fatal Unk

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h) T cell and B cell profile after splenectomy

T cell profile Date
Day Month Year

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CD8 CD27+ CD28-	CD8 effector T cells	<input style="width: 60px;" type="text"/>	<input style="width: 60px;" type="text"/>	<input type="radio"/>

B cell profile Date
Day Month Year

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