

**Table S1.** Personal data of APS1 patients reported until 1980. Data given in red indicate subclinical diseases. The asterisk indicates probable decease by acute adrenal failure

Author Year	Gender	Initials	Birth Date	CMC	HPT	AAD	GI	AG	T1D	ATD	M	AH	K	A	V	EH	Last Obs.	Death †	Autopsy – Pathologic changes of adrenal cortex – Histology
Thorpe and Handley 1929 <i>Philadelphia (PA), USA</i>	F	EP	Oct 01, 1921	4	4			5			5					7 †		Not performed	
Ostertag 1930 <i>Berlin, Germany</i>	F	CL			1	2										2 †*	Atrophy and fibrosis – Lymphocyte and mononuclear cell infiltrate		
Söderlund 1938 <i>Helsinki, Finland</i>	M	MH			+		+									21			
Emerson et al. 1941 (updated by Gass 1962) <i>Baltimore (MD), USA</i>	M	JH		+	13						4	+		+	18 †		Undisclosed		
McQuarrie et al. 1941 (updated by Papadatos and Klein 1954, and DiGeorge and Paschkis 1957) – <i>Minneapolis (MN), USA</i>	M	LW		13	19									+	19 †	Complete atrophy – Histology not reported			
Sevinghaus 1942 (updated by Sutphin et al. 1943) <i>Madison (WI), USA</i>	F	DE		4	5										27				
Sutphin et al. 1943 (updated by Kantrowitz et al. 1969) <i>Boston (MA), USA</i>	M▲ AG			0	10										35				
	F▲ LG			2	5										14 †	Not performed			
	F▲ LG			4	11	12								+	12 †*	Not performed			
	F ER			1	7										7				
	F SAH			+	3									+	7				
Talbot et al. 1943 (updated by Talbot et al. 1954, Craig et al. 1955, and Fisher and Fitzpatrick 1970) – <i>Boston (MA), USA</i>	F▲			+	4	8									10 †*	Not performed			
	M▲		Sep 26, 1936	+	6	6								+	6 †	Atrophy – Lymphocyte and plasma cell infiltrate			
	F▲			0	6	14								14	21				
Leonard 1946 <i>New York (NY), USA</i>	F	EK		3	12						10	+		+	12 †*	Severe atrophy and fibrosis – Lymphocyte infiltrate			
Keating 1947 (updated by Lovestedt 1950, and Hermans et al. 1969) – <i>Rochester (MN), USA</i>	F▲			2	6						2	+	7	+	45				
	F▲			3	18	13								+	19 †*	Undisclosed			
Gotta and Odoriz 1948 <i>Buenos Aires, Argentina</i>	M	AC		+	16										21				
Berezin and Stein 1948 (updated by Weinberg et al. 1976) <i>New York (NY), USA</i>	M▲ ER		Mar 08, 1940	7	4	33	33	33	33		20	28	+	34					
	M▲				+										?				
Collins-Williams 1950 (updated by Craig et al. 1955) <i>Boston (MA), USA</i>	M▲		Apr 28, 1940	1	6	6			2		7				11 †*	Severe atrophy – Lymphocyte and plasma cell infiltrate			
	M▲ LW			1	6		6		6	4	4				14				
Author Year	Gender	Initials	Birth Date	CMC	HPT	AAD	GI	AG	T1D	ATD	M	AH	K	A	V	EH	Last Obs.	Death †	Autopsy – Pathologic changes of adrenal cortex – Histology
Salvesen and Bøe 1953 <i>Oslo, Norway</i>	M	LM	Mar 17, 1940	0	7	8					7					8 †		Undisclosed	
Leifer and Hollander 1953 <i>New York (NY), USA</i>	M	PH			8	19					8					20			
McLean 1954 <i>Aberdeen, United Kingdom</i>	F			+	4										11				

Papadatos and Klein 1954 <i>Pittsburgh (PA), USA</i>	M	YS		10	10		9	5	+	10			
Blackburn 1954 <i>Sidney, Australia</i>	F			10	23					31			
Castleman and Towne 1954 (updated by Dudley et al. 1955) <i>Boston (MA), USA</i>	F	JG		2	3	4			4 †*	Severe atrophy – Lymphocyte and plasma cell infiltrate			
Craig et al. 1955 <i>Boston (MA), USA</i>	F		Jun 14, 1941	3	7	7		7	12 †*	Complete atrophy and fibrosis – Mononuclear cell infiltrate			
Perlmutter et al. 1956 <i>New York (NY), USA</i>	F	JL		10	27	24	+	+	+	30 †	Severe atrophy and fibrosis – Lymphocyte infiltrate		
Forbes 1956 <i>Rochester (NY), USA</i>	F	LJH	Nov 11, 1946	4	5			1		8			
	F	EF	Jul 21, 1941	1	11				+	13 †*	Complete atrophy and fibrosis		
	F	KW	Feb 14, 1948	2	4			3	+	6			
Sano et al. 1956 <i>Sendai, Japan</i>	F			3	4	5			+	6			
Whitaker et al. 1956 <i>Middletown (OH), USA</i>	M	NA		2	15	15	14	+	15	†	Severe atrophy – Lymphocyte infiltrate		
DiGeorge and Paschkis 1957 <i>Philadelphia (PA), USA</i>	?▲				5				5 †	Undisclosed			
	?▲				10	8				10			
	?▲			3	6			2		6			
Wilkins 1957 (updated by Gass 1962) <i>Baltimore (MD), USA</i>	F	AO'M	Jul 12, 1940	+	4		12	3	0	5	+	20	
Malloy and Woodruff 1958 <i>Nashville (TN), USA</i>	M			+	1	5		6		7 †	Not performed		
Järvinen and Latvalahti 1958 (updated by Siurala et al. 1968) <i>Helsinki, Finland</i>	F▲	EE		10	5		32	37	4	10	45		
	M▲				0					0 †	Undisclosed		
	F▲				+					3 †	Undisclosed		
	M▲			10				+	10		35		
Hetzl and Robson 1958 <i>Adelaide, Australia</i>	F			0	10	15	13		12	+	17		
Fargašová 1958 (updated by Fárová 1971) <i>Olomouc, former Czechoslovakia</i>	F	VL	..., 1945	+	8	11			8	+	16 †*	Severe atrophy – Histology not reported	
Akers et al. 1958 (complemented by Irvine et al. 1968, and updated by Kolb et al. 1970) – <i>Denver (CO), USA</i>	F	MH	Aug .., 1948	1	12	13		2			20		
Cathala et al. 1958 (updated by Brun 1975, and Cortet et al. 1978) – <i>Dijon, France</i>	F	MFC	Apr 29, 1954	2	2	17	13	18			23		
Svane-Knudsen 1959 (updated by Axelsen 1971) <i>Kolding, Denmark</i>	F▲			32	7		+		26		56		
	F▲				+						21 †	Undisclosed	
Cramblett et al. 1959 <i>Iowa City (IA), USA</i>	F▲			4	7						7		
	M▲			+	+						? †*	Undisclosed	
McMahon et al. 1959 <i>Madison (WI), USA</i>	F			+	6	19	13		6	7	+	21 †	Severe atrophy – Mononuclear cell infiltrate
Williams and Wood 1959 <i>London, United Kingdom</i>	F				6	7			6	7	+	7 †	Focal atrophy – Lymphocyte infiltrate
Szczepańska and Sapiecha 1959 <i>Warsaw, Poland</i>	F	ZB	..., 1952	+	5					6 †	Extreme atrophy – Histology not reported		
Anning 1959 <i>Leeds, United Kingdom</i>	M			+	0						14		
Prader et al. 1959 <i>Zurich, Switzerland</i>	M	JCP	Mar 22, 1944	+	12			3		12 †	Atrophy and fibrosis – Lymphocyte infiltrate		
Carter et al. 1959 <i>New York (NY), USA</i>	F	CV		3	4		+		5	6 †	Severe atrophy – Lymphocyte infiltrate		
Chaptal et al. 1960 (updated by Vayssettes-Vernet 1976) <i>Montpellier, France</i>	F▲	EC	Jun 30, 1953	2	3	9				18 †	Undetectable gland		
	F▲	SC	Nov 26, 1956	0	3	14		0		14 †	Atrophy – Lymphocyte infiltrate		



Galusha and Kelly 1965 <i>Charlotte (NC), USA</i>	F▲ F▲	0 3 0	+ +	17 0 †* Undisclosed
Marchenko et al. 1965 <i>Moscow, former USSR</i>	F OI	1 3 4 13	0	+ 4
Sjöberg 1966 <i>Mölndal, Sweden</i>	M▲ F▲ F	6 14 2 18 + 12 22	14	+ 18 + 25 + + 22
Sweetnam 1966 <i>Huddersfield, United Kingdom</i>	F▲ F▲	7 7 4 4		7 4
Pisanty 1966 (updated by Moshkowitz et al. 1969, and Pisanty and Garfunkel 1977) – <i>Jerusalem, Israel</i>	F MH	2 0	8	+ + 10
Conte et al. 1967 (updated by Panizon 1969) <i>Padua, Italy</i>	F▲ GC ... , 1950 F▲ ALC May., 1951 F▲ EC ... , 1955	6 13 0 8 17 13 + 3 10	10 0 4 + + + 0 4 +	18 17 13
Wuepper and Fudenberg 1967 (updated by Kirkpatrick et al. 1971) – <i>San Francisco (CA), USA</i>	M GR	2 19 14 20 + 20 + 18 19	+ 24	†* Undisclosed
Jackson and Whyte 1967 <i>Glasgow, United Kingdom</i>	M	17 25		29
Brodehl et al. 1967 <i>Bonn, former West Germany</i>	F▲ KK ... , 1961 M▲ M▲	+ + 9 0		4 9 † Undisclosed 0 † Undisclosed
Chilgren et al. 1967 <i>Minneapolis (MN), USA</i>	M M▲ F▲ F▲	13 17 0 14 9 9 9 8 8		23 25 17 13
Mariani et al. 1967 (updated by Mariani et al. 1971) <i>Marseille, France</i>	M▲ JPT F▲ BT	5 8 6 6		8 †* Undisclosed 6
Suchnicka 1967 <i>Wroclaw, Poland</i>	F	7 16 15		16
Austoni et al. 1968 (updated by Del Prete et al. 1975) <i>Padua, Italy</i>	F MTF	+ 7 18 21 25 25		25
Sethi et al. 1968 <i>New Dehli, India</i>	M MN	+ 8	+ 14	
Golonka and Goodman 1968 <i>Albany (NY), USA</i>	F	+ 3 17 13		20
Vukadinović et al. 1968 <i>Zagreb, former Yugoslavia</i>	F BT	6 7 + 6 2		12
Panizon 1969 <i>Trieste, Italy</i>	F PH	Dec 19, 1964 0 2	3	4
Hermans et al. 1969 <i>Rochester (MN), USA</i>	F	4 29 22	21	33 † Severe atrophy – Lymphocyte infiltrate
Quichaud et al. 1969 (complemented by Massy 1969) <i>Amiens, France</i>	M▲ JBH F▲ MH	9 9 9 0 0	9	+ 13 4 † Undisclosed
Clarke 1969 <i>Melbourne, Australia</i>	M ST	2 10 16		19
Riley 1969 <i>Danville (PA), USA</i>	F	+ 6	+ +	13
Goudie et al. 1969 <i>Glasgow, United Kingdom</i>	M	9 3 22	19	+ 22
Haddock and Blizzard (reported by Irvine 1969 and complemented by Irvine et al. 1969) – <i>San Juan (PR), USA</i>	F	2 19		20
Alteraş et al. 1969 <i>Bucharest, Romania</i>	F	+ 14 13		14
Greenberg et al. 1969 <i>Greenberg et al. 1969</i>	M	0 11	7 + +	17

<i>Philadelphia (PA), USA</i>	M	DD	... ., 1955	13	13	12	14		+	14
Peillon et al. 1970 <i>Reims, France</i>										
Meehan 1970 (updated by Mackey 1975) <i>Dublin, Ireland</i>	M		... ., 1954	0	18	+ 17				18
Kirkpatrick et al. 1970 (updated by Kirkpatrick et al. 1971) <i>Kansas City (KS), USA</i>	M▲	WM		2	8		17			18
	M▲	RM		3	8		7			16
Drury 1970 (complemented by Irvine et al. 1968) <i>Dublin, Ireland</i>	F▲	ML	Jan 12, 1945	7	16	15	13	21	15	5 24 †* Undetectable gland
Krebs and Kuske 1970 <i>Bern, Switzerland</i>	F▲	RA	... ., 1962	2	2				6	+ 8
	M▲	BA	... ., 1966	0	4				2	4
Foz et al. 1970 (updated by Foz et al. 1973) <i>Barcelona, Spain</i>	F▲	AAP		24	22					26
	F▲	EAP		+	12					14
Windorfer 1970 <i>Freiburg in Brisgow, former West Germany</i>	M	WL	Jun 12, 1959	0	9	10		2		10
Nally 1970 <i>Dublin, Ireland</i>	F▲			0	2	13			+	+ 22
	M▲			6	17	17				17
Castanier et al. 1970 <i>Metz, France</i>	F		Sep 19, 1964	0	4					5
Ferchiou et al. 1970 <i>Tunis, Tunisia</i>	M			0	11					11
Irvine 1970 (complemented by Irvine et al. 1968) <i>London, United Kingdom</i>	F			19	13	18			19	26

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Block et al. 1971 <i>Chicago (IL), USA</i>	F	JW		22	7	23	16										29		
Fields et al. 1971 <i>Concord (MA), USA</i>	M			2	14						14	14					15		
Blizzard 1971 <i>Baltimore (MD), USA</i>	F	SW	... ., 1955	1	14				+	+							14		
Fárová 1971 <i>Olomouc, former Czechoslovakia</i>	F	HH	... ., 1952	+	5	13					5	+					16 †* Undisclosed		
	M	JL	... ., 1955	+	10							+					16		
Money et al. 1971 <i>Baltimore (MD), USA</i>	M▲		Oct 23, 1949	3	5	5		0	+	16							19		
	M▲		Mar 03, 1951	8	4	7		16	4	+		7					18		
	M▲		Sep 27, 1956	2	7	0											12		
Castells et al. 1971 <i>New York (NY), USA</i>	F▲	MC		6	7	7									+	7			
	M▲	MC		+		6										6			
	F▲	MC		+		4										4			
Levy et al. 1971 (updated by Arvanitakis and Knouss 1973) <i>Madison (WI), USA</i>	M			3	8	+		+								17			
Kössling and Emmrich 1971 <i>Mainz, former West Germany</i>	M			2	11											12 †* Complete atrophy and fibrosis – Lymphocyte infiltrate			
Stankler and Bewsher 1972 <i>Aberdeen, United Kingdom</i>	F▲	HC	... ., 1951	+	15	13			3	3						18			
	F▲	KC	... ., 1951	1	18											18			
	M▲	DC	... ., 1955	8	14											14			
Témime 1972 <i>Marseille, France</i>	F			5	10											11			
Olin and Poindexter 1972 <i>Fargo (ND), USA</i>	F▲		Oct 30, 1951	+	6	19	11								+	19			
	F▲		May 11, 1953	10	4										+	17			

Rameis et al. 1972 <i>Innsbruck, Austria</i>	M▲ HO	May 27, 1953	3				+ 18
	F▲ EO	Feb 03, 1955	3	3	13	12	+ 16
	F IB	Oct 02, 1967	1	0			3
	F SF	Aug 19, 1962	5	3	5	1	7
Breynaert et al. 1973 (complemented by Dumortier-Iscovich 1973) – <i>Lille, France</i>	M▲ TF	Jun 28, 1962	5	7		2	10
	M▲ BF	Mar 25, 1957	0		14	10	16
Mitschke et al. 1973 <i>Hamburg, former West Germany</i>	F		5	7	7	5	7 † Severe atrophy and fibrosis – Lymphocyte and macrophage infiltrate
Arvanitakis and Knouss 1973 <i>Madison (WI), USA</i>	M▲		7	14	23	+ 23	+ 23
	M▲		5		8		8 †* Undisclosed
Scott 1973 <i>Craigavon, United Kingdom</i>	F			16	+		16
Hooper et al. 1973 <i>Sidney, Australia</i>	M▲			3			24
	M▲				12		12
Corvaglia 1973 <i>Florence, Italy</i>	F▲ LM		+ 2	5			5
	F▲ AM		+ 6	8			8
Wong and Kirkpatrick 1973 (complemented by Kirkpatrick et al. 1971) – <i>Washington (DC), USA</i>	M RH		0	7		+ +	17
Langslet et al. 1974 <i>Oslo, Norway</i>	M		+ + +				7
Marieb et al. 1974 <i>Boston (MA), USA</i>	F		+ 12	13		12	13
Kleerekoper et al. 1974 <i>Sidney, Australia</i>	F	Oct 28, 1956	13	5	16	8	+ 17
Krohn et al. 1974 (updated by Mylläniemi and Perheentupa 1978) – <i>Helsinki, Finland</i>	F RH	..., 1959	12	12	12		+ 14
	F VT	..., 1965	1	1	6		+ 10
	F LP	..., 1961	6	6	12	+ 6	12
	F LR	..., 1965	1	2	8	1	+ 11
	M▲ SA	..., 1961	2		9	7	+ 14
	F KL	..., 1963	1	3	9	9	+ 12
	M TW		8	8		10	10 † Undisclosed
	M▼ TM	..., 1963	8	1			+ 12
	F▼ NM	..., 1967	2	1	5		+ 8
	M▼ KM	..., 1961	3	4	4		+ 14
	F MK		1	11			11
	F HL	..., 1967	3	3			+ 8
	F AN	..., 1966	2	6		2	8
	M▲ PA	..., 1966	1	6			10
Conte et al. 1974 <i>Padua, Italy</i>	F▲ EB	..., 1929	33	26			43
	M▲ EB	..., 1934	+ 36				38
Jehanne and Guivarch 1974 <i>Roscoff, France</i>	F GB	Mar 10, 1955	2	2	13	0	10
Bovier-Lapierre and Jeune 1974 <i>Lyon, France</i>	M DP		+ 3	3		5	5 † Undisclosed
Schulkind and Ayoub 1975 <i>Gainesville (FL), USA</i>	F		1	3	+	3	12
Kaffe et al. 1975 <i>New York (NY), USA</i>	F▲ MC		7	7			12
	M▲ MC		+ 10				+ 10
	F▲ MC		4	8		7	8
Ellwood et al. (reported by Irvine and Barnes 1975) <i>Bristol, United Kingdom</i>	F▲		+ 9	10	13	8 + 15	15
	M▲		7			7 0	13
Joksimović et al. 1975 <i>Belgrade, former Yugoslavia</i>	M▲ RS	Jun 17, 1956	2			3 2	3 † Not reported
	F▲ GS	Dec 11, 1960	3	4		4 6	6 † Not performed
	F▲ SS	Sep 09, 1964	2	3		1 3	3 †* Severe atrophy – Lymphocyte infiltrate

	M▲ VS	Feb 05, 1969	2	1	2	
	F BB	Apr 04, 1964	4	3 5	4 4 4	6 †* Not performed
	M RT	Jun 12, 1954	7	7	12 12	19
Petković 1975	M		5	15	7 7	23
Zagreb, former Yugoslavia						
Champenois 1975	F NP	Mar 29, 1969	3	3	3	6
Lille, France						
Heinonen et al. 1976	M JK	..., 1946	0	19 20	17	26
Tampere, Finland						
Lawton et al. 1976 (updated by Kessel and Taylor 1980)	M▲ KH	Aug 14, 1968	0	4		9
Edinburgh, United Kingdom	M▲ MH	Oct 11, 1975	0			2
Chesney et al. 1976	M PR		3 13 14		6	16
Montreal, Canada						
Matsumoto et al. 1976	M		+ + +			16
Fukuoka, Japan	F		4 12			14
Farrell et al. 1976	F		2 6 9			9
Madison (WI), USA						
Blake 1976	F		+ 3 18			25
Dublin, Ireland	F		+ 18		+	22
Vayssettes-Vernet 1976	F PP	Dec 04, 1962	11 11 11 13			13
Grenoble, France						
Stieglitz et al. 1977	F▲		2	4	+	15
Toronto, Canada	F		+	9	+	17
	M		+ + +		+	10
	M		+ + +		+	6
	M▲		+ +			19
	F		+ +			5
Lucky et al. 1977	F		0 4 11	0	4	17
Bethesda (MD), USA	F▲		7 8 14			14
	M▲		+ + +	+	+	11
Hertz et al. 1977	F		6 9 34 15 13		6 28	34
Bethesda (MD), USA	F		3 6 10 16		3 3	18
Chesney et al. 1977	F	..., 1962	12 7	+ 7		14
Madison (WI), USA						
Pisanty and Garfunkel 1977	F▲ VA		9 18	+	+	18
Jerusalem, Israel	M▲ IA		8		+	21
	F ZS		+ 17		+	18
	M IA		+ +		+	17
	M▼ NS		+ 7		+	30
	M▼		+ +		+	?
Rebollar et al. 1977	F		+ 21			34
Madrid, Spain						
Miani and Rumi 1977	M FG		6 + +			14
Rome, Italy						
Dučić and Terzić-Kazić 1978	M▲ BM		+ 8 11		+	11
Sarajevo, former Yugoslavia	F▲		+			9
Prader 1978	M▲		+	+		3 † Undisclosed
Zurich, Switzerland	M▲		+ +	+		7 † Undisclosed
	M▲		+ +			3 † Atrophy – Histology not reported
	M▲		2	2		8 † Undisclosed
Myläniemi and Perheentupa 1978	F MH		0 4		+	5
Helsinki, Finland	F AR		10 5 10		+	11
	F MK		6 7		+	8
	F MP		3 3 9		+	14

	F	SK	7	23	23		+	23	
	M	MP	5	8			+	11	
	M	HK	10	10	10		+	17	
	M	KK	7	18			+	22	
Brenton et al. 1978 <i>Colchester, United Kingdom</i>	M	... ..., 1940	+	34		+		36	
Tomar et al. 1979 <i>Syracuse (NY), USA</i>	F	... ..., 1955	10	2	20	13	17	10	20
Betterle et al. 1979 <i>Padua, Italy</i>	F		+	+	+	+		+	25
Matzner et al. 1979 <i>Jerusalem, Israel</i>	F		3	15	15		10	16	
Arulanantham et al. 1979 <i>New Haven (CT), USA</i>	M▲		7	7		+	3	12	
	F▲		5	4	4			6	
	F▲		2		2		2	3 † Not reported	
Ivanova and Ignatovich 1979 <i>Omsk, former USSR</i>	M		2	5		3	+	9	
Myers et al. 1980 <i>Hartford (CT), USA</i>	F▲		2	15	13			26	
	M▲			6			6 †*	Undisclosed	
Adami Lami et al. 1980 <i>Florence, Italy</i>	F▲ CS		0	8	13	13	+	13	
	M▲		+					13	
Clifton-Bligh et al. 1980 <i>Sidney, Australia</i>	M	DOB	Jul 21, 1952	0	10	22	+	25	
Rico Zalba and Calderin 1980 <i>Madrid, Spain</i>	M▲			+			? †*	Undisclosed	
	M▲ DVC		+	8	14		+	14	
	F▲ FVC		+	11		2		19	
	M▲ SVC			13				14	

CMC, chronic mucocutaneous candidiasis; HPT, idiopathic hypoparathyroidism; AAD, autoimmune Addison's disease; GI, gonadal insufficiency; AG, atrophic gastritis; T1D, type-1 diabetes; ATD, autoimmune thyroid disease; M, malabsorption; AH, autoimmune hepatitis; K, keratitis; A, alopecia; V, vitiligo; EH, enamel hypoplasia

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