

Since this is a very extensive table, the format and content has not been edited by ActaDV.

**Table SI.** Characteristics of 38 reported cases of Schöpf-Schulz-Passarge Syndrome

Author	Year	N	Sex	Age	Cysts	Since age	Hypodontia	1° dentition	2° dentition	Comment	Hypotrichosis	Since age	Palmo-plantar hyperkeratosis	Since age	Sweating	Since age	Onychodystrophy	Since age	Consanguinity	Pedigree presented	Malignant tumors	At age	
Schöpf et al.	1971	2	F	68	X	60	X	Lost early	X	Only 3 permanent teeth (12, 11, 48)	X	25	X	12			X	12	Yes	X			
			F	66	X	Late in life	X	X	X		Yes		X										
Burket et al.	1984	1	M	35	X	30	X	N.a.	X	Only 4 (rudimentary) permanent molars, persistent deciduous teeth until his late 20s, no hx of periodontitis Fewer permanent teeth, most of them widely spaced, no hx of periodontitis			X	12			X	12	No				
Font et al.	1986	1	M	67	X	20	X				X	20			X				No		BCC nose	66	
Nordin et al.	1988	3	F	80	X	65	X	X	X	4 lower deciduous teeth missing, lost at age 8-9, only 4 cone-shaped upper incisors (12, 11, 21, 22) developed Small deciduous teeth, 2 lower permanent incisors missing, 2 upper permanent incisors cone-shaped, other permanent teeth small and widely spaced	X		X	15	X	Always	X	Childhood	No	X			
			M	75	X	60	X				20	X	15	X	Youth	X	Childhood						
Perret et al.	1989	1	M	75	X		X		X	Denture at age 18 because of hypoplastic teeth	X	20	X				X			BCC? (clinical diagnosis) SSC	71 67, 75		
Monk et al.	1992	3	F	62	X		X	X	X	Primary dentition reported "abnormal", no permanent teeth, edentulous at exam; "bird-like facies" "unusual" primary dentition, no permanent dentition, edentulous at exam; "bird-like facies" One single permanent tooth only, edentulous at exam, "bird-like facies"	X		X	47			X		Yes		Hypernephroma, BCC		
			M	58	X		X	X	X		X								Yes				
			M	51	X		X		X		X	X							X	Yes			
Küster & hammerstein	1992	1	F	43	X	43	X	N.a.	X	Most permanent teeth missing, first dentures at age 14, 5 permanent teeth present at exam	X	Childhood	X	20			X		No	X			
Craigen et al.	1997	1	M	56	X	54	X		X	No permanent teeth, total dental extraction at age 18	X	Life-long	X	Life-long			X	Life-long	No	X			
Starink	1997	4	F	78	X	77				Small conical widely spaced teeth At age 16 all teeth were removed, because of conical shape Conical teeth Only part of the permanent teeth developed	X		X	23			X		No		Malignant ESFA		
			F	59	X				X			X	20						No				
			F	73	X						X		Childhood						X	No		BCC of nose, adenocarcinoma of breast	
			F	71	X		X		X		X	X	25						X	No			
Verplancke et al.	1998	1	M	53	X		X	N.a.	X	No permanent teeth developed	X	18	X	14			X		No				
Dot et al.	2000	1	M	71	X	55	X	X	X	Missing primary incisors and conical canines; only 8 permanent teeth, conical upper permanent canines	X	30	X	20			X	45	No		BCC, M. Bowen	45 and 70	
Gkolfinopoulos et al.	2001	2	F	49	X		X	N.a.	X	Permanent teeth did not erupt, "bird-like facies", full dentures Permanent teeth did not erupt (atrophic roots on OPG), full dentures	X		X	25			X		No		No		
			M	56	X		X		X		X	30	X	18	X	Child-hood	X	Yes		No			

Hampton et al.	2005	1	M	69	X	50	X			Full dental clearance @ age 14	X	15	X	20		X	10			
Castori et al.	2008	2	F	65	X	25 (?)	X	N.a.	X	Only 8 permanent teeth, "bird-like appearance"	X	Childhood	X	Childhood		X	Childhood	Yes	X	
			M	73	X	50	X	N.a.	X	Complete absence of permanent dentition	X	Early childhood	X	43	X	Adult-hood	X	Early childhood	Yes	X
Bohring et al.	2009	1	F	25	X		X	X	X	17 permanent teeth missing (including wisdom teeth), 12 persistent deciduous teeth, 3 permanent teeth present	X		X		X				Porocarcinoma of left heel	
Szepetiuk	2009	1	M	56	X	46	X			"bird-like appearance", dental alterations present "at birth", majority of permanent teeth absent	X		X	15		X	Birth	No	PMH: several SSC, BCC, s/p cardiac transplantation in 1992, med:immunosuppression	
Nagy et al.	2010	1	F	70	X	40	X	X	X	Reduced no. Of deciduous teeth, no permanent teeth	X	Childhood			X		X	Early adulthood	No	
Petrof et al.	2011	1	F	59	X	50	X	X	X	Reduced primary dentition, lack of secondary dentition			X	30-40	X	30-40	X	Unknown	Fostered child	
Wedgeworth et al.	2011	1	M		X		X		X	Severe hypodontia, dentures from the age of 14, conical-shaped teeth								No	Malignant melanoma, cutaneous SCC, BCC	
Tziotzios et al.	2014	7	F	58	X		X	X	X		X		X		X		X			
			M	50	X		X	X	X		X		X		X		X			
			M	43	X		X	X	X		X		X		X		X			
			F	36	X		X	X	X		X		X		X		X		BCC	
			M	57	X		X	X	X		X		X		X		X		Basosquamous CA	
			M	28	X		X	X	X		X		X		X		X			
			F	49	X		X	X	X		X		X		X		X			
Vilas-Sueiro et al.	2015	1	M	70	X	Many years	X		X	No 2nd teeth since childhood	X		X		X		X			
Painsi et al.	2017	1	F	86	X		X	X	X		X		X	Life-long	X	Childhood	X	No	Squamous cell CA of right 50 palm excised	
<b>Total</b>		<b>38</b>																		
<b>Mean</b>				<b>59,44</b>								<b>22</b>		<b>22,1</b>						
<b>Min</b>												<b>15</b>		<b>12</b>						
<b>Max</b>												<b>30</b>		<b>47</b>						
<b>Median</b>												<b>20</b>		<b>20</b>						
<b>Present study</b>	<b>2018</b>	<b>4</b>	<b>M</b>	<b>53</b>	<b>X</b>		<b>X</b>	<b>N.a.</b>	<b>x</b>	<b>Only 6 permanent teeth, 2 persisting deciduous teeth</b>	<b>X</b>	<b>17</b>	<b>X</b>	<b>20</b>	<b>N.a.</b>	<b>X</b>		<b>no</b>	<b>x</b>	
			<b>F</b>	<b>51</b>	<b>X</b>		<b>X</b>		<b>X</b>	<b>Only 8 permanent teeth</b>	<b>X</b>		<b>(x)</b>		<b>Almost no sweating</b>	<b>X</b>	<b>50</b>	<b>No</b>	<b>X</b>	
N.a. = no abnormalities			<b>F</b>	<b>46</b>	<b>X</b>								<b>X</b>					<b>No</b>	<b>X</b>	
			<b>F</b>	<b>25</b>			<b>X</b>		<b>X</b>	<b>Agenesis of 5 permanent teeth</b>			<b>X</b>					<b>No</b>	<b>X</b>	
			<b>F</b>	<b>8</b>			<b>X</b>		<b>X</b>	<b>Agenesis of 5 permanent teeth</b>			<b>X</b>					<b>No</b>	<b>X</b>	