

Table SI. ICD-10 codes for infections and hyperthermia in the Danish National Patient Registry

Category	ICD-10 codes
Hospital-treated infections	
Any infection	A00-A99, B00-B49, B95-B99, D73.3, E06.0, E06.9, E32.1, G00-G02, G04-G07, H00, H01.0, H03.1, H04.0, H04.3, H05.0, H10, H13.0-1, H15.0, H19.1-2, H22.0, H32.0, H44.0-1, H60.0-1, H60.3, H62.0-3, H65-H67, H68.0, H70.0, H70.2, H73.0, H75.0, H94.0, I30.1, I32.0, I33.0, I38, I39.8, I40.0, I41, I43.0, I52.0-1, I68.1, I98.0-1, J00-J06, J10-J18, J20-22, J34.0, J36, J38.3D, J38.7G, J39.0-1, J39.8A, J44.0, J85.1-3, J86, K02, K04.0, K04.6-7, K05.2, K11.2-3, K12.1-2, K13.0A, K13.0C, K14.0A, K20.9A, K23.0, K35, K37, K57.0, K57.2A, K57.4, K57.8, K61, K63.0, K65.0, K65.8I, K67, K75.0-1, K77.0, K80.0, K80.3-4, K81.0, K83.0, K85.8A, K85.8E, K85.9, K93.0-1, L00-08, M00-01, M46.1-5, M49.0-3, M60.0, M60.8, M63.0-2, M65.0-1, M68.0, M71.0-1, M86.0-2, M86.9, M90.0-2, N10, N12, N13.6, N15.1, N15.9, N16.0, N20.0I, N29.1, N30.0, N30.8, N33.0, N34.0-1, N39.0, N41, N43.1, N45.0, N45.9, N48.1-2, N49, N61, N70-77, O23, O26.4, O41.1, O75.3, O85, O86, O91, O98, P35-39, T80.2, T81.4, T82.6-7, T83.5-6, T84.5-7, T85.7, T88.0, T89
By selected categories:	
Gastrointestinal tract infections	A00-A09
Abscesses	A06.5, A54.1A, B43.1-2, D73.3, E06.0A, E32.1, G06, G07, H00.0A, H05.0A, H44.0A, H60.0, J34.0A, J36, J38.3D, J38.7G, J39.0-1, J39.8A, J85.1-3, K04.6-7, K11.3, K12.2, K13.0A, K14.0A, K20.9A, K35.3A, K35.3B, K57.0, K57.2A, K57.4, K57.8, K61, K63.0, K65.0A-L, K75.0, K81.0A, K85.8A, L02, L05.0, L05.9, M60.8A, M86.9A, N15.1, N15.9, N34.0, N41.2, N45.0, N48.2, N49.2A, N61.9A, N61.9B, N70.0A, N70.0B, N71.0A, N73.0A, N73.0B, N73.2A, N73.2B, N73.3A, N73.5A, N73.8A, N73.8C, N75.1, N76.4, N76.8A

Septicaemia	A40, A41
Skin and subcutaneous infections	A46, B00-B09, J34.0, L00-08
Viral infections	A80-A99, B00-B09, B15-B19, B25-B34, B97, J10-12, I41.1
Fungal infections	B35-B49
Eye and ear infections	H00, H01.0, H03.0-1, H04.0, H04.3, H05.0, H06.1, H10, H13.0-1, H15.0, H19.1-2, H22.0, H32.0, H44.0-1, H60.0-1, H60.3, H62.0-3, H65-H67, H68.0, H70.0, H70.2, H73.0, H75.0, H94.0
Pneumonia	J10.0, J11.0, J12-18, J22
Upper respiratory tract infections	J00-06, J36, J38.3D, J38.7G, J39.0-1, J39.8A, K04.0, K04.6-7, K05.2, K11.2-3, K12.2, K13.0A, K14.0A
Teeth and oral cavity infections	K02, K04.0, K04.6-7, K05.2, K11.2-3, K12.1-2, K13.0A, K13.0C, K14.0A, B00.2
Intraabdominal infections	K20.9A, K23.0-1, K35, K37, K57.0, K57.2, K57.4, K57.8, K61, K63.0, K65.0, K65.8I, K67, K75.0-1, K77.0, K80.0, K80.3-4, K81.0, K83.0, K85.8A, K85.8E, K85.9, K93.0-1
Urinary tract infections	N10, N12, N15.1, N30.0, N30.8, N33.0, N34.0-1, N39.0
Hyperthermia / pyrexia:	
Hyperthermia	P81.8, P81.9, R50 (excluding R50.2), R50.8, R50.9
Febrile convulsions	R56.0

Codes to define infectious disease categories were modified from Mor *et al.* (1)

Abbreviations: ICD, International Classification of Diseases

Table SII. ATC codes for antimicrobial prescriptions in the National Prescription Registry

Category	ATC codes
Systemic antibiotics, any	J01
Selected subgroups:	
Phenoxymethylpenicillin	J01CE02
Aminopenicillins: pivampicillin, amoxicillin, amoxicillin with β -lactamase inhibitor	J01CA02, J01CA04, J01CR02
Macrolides: azithromycin, erythromycin, roxithromycin, clarithromycin	J01FA10, J01FA01, J01FA06, J01FA09
Cystitis antibiotics: pivmecillinam, sulfamethizole, nitrofurantoin, trimethoprim	J01CA08, J01EB02, J01XE01, J01EA01
Beta-lactamase resistant penicillins: dicloxacillin, flucloxacillin	J01CF01, J01CF05
Quinolones	J01M
Systemic antimycotics	J02
Systemic antivirals	J05
Ophthalmic antimicrobials	S01A, S03A, S03C
Otological antimicrobials	S02A, S03A, S03C

Abbreviation: ATC, Anatomical Therapeutic Chemical classification system

Table SIII. Adjusted analysis of all-cause mortality and cause-specific mortality

Subgroups	Patients with ED			Matched comparators			HR (95% CI)	
	Events, n/total	Person- years at risk	Mortality rate per 100,000 person years	Events, n/total	Person- years at risk	Mortality rate per 100,000 person years	Unadjusted ^a	Adjusted ^b
Any ED	11/396	4995	220	71/3960	50153	142	1.33 (0.70–2.55)	1.44 (0.75–2.77)
Males	7/150	2061	340	19/1500	20922	91	3.77 (1.57–9.03)	4.58 (1.74–12.07)
Females	4/246	2934	136	52/2460	29231	178	0.59 (0.21–1.68)	0.62 (0.22–1.77)
Hypohidrosis males	5/78	1259	397	12/780	12905	93	4.39 (1.53–12.65)	4.33 (1.20–15.60)
Hypohidrosis females	<3/57	N/A	N/A	16/570	6998	229	N/A	N/A
Diagnosed <18y old	4/244	3192	125	7/2440	32695	21	6.53 (1.84–23.13)	8.36 (2.08–33.70)
Diagnosed ≥18y old	7/152	1803	388	64/1520	17458	367	0.89 (0.40–1.98)	1.00 (0.44–2.23)
Males with XLHED	<3/50	N/A	N/A	7/500	8338	84	N/A	N/A
<i>Cause-specific mortality related to infectious and respiratory disease</i>								
Any ED	5/396	4627	108	17/3960	46459	37	2.20 (0.78–6.17)	3.05 (0.98–9.52)

^aAdjusted for matching factors (age, sex, and municipality) by design.

^bAdjusted for educational level

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; IP, incontinentia pigmenti; HR, hazard ratio; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SIV. Infections and hyperthermia before ectodermal dysplasia diagnosis

Subgroup	ED patients exposed ^a , n (%)	Comparators exposed, n (%)	Unadjusted ^b OR ^c (95% CI)	Adjusted ^d OR ^c (95% CI)
Any ED (n=396)				
Any infection	183 (46.2)	1163 (29.4)	2.27 (1.81–2.85)	2.26 (1.80–2.84)
GI tract infections	32 (8.1)	161 (4.1)	2.11 (1.41–3.16)	2.09 (1.40–3.12)
Abscesses	14 (3.5)	112 (2.8)	1.27 (0.71–2.26)	1.27 (0.71–2.26)
Septicaemia	6 (1.5)	14 (0.4)	4.29 (1.65–11.15)	4.17 (1.60–10.87)
Skin and SC infections	36 (9.1)	170 (4.3)	2.29 (1.56–3.36)	2.29 (1.56–3.37)
Viral infections	50 (12.6)	206 (5.2)	2.76 (1.96–3.87)	2.72 (1.94–3.82)
Fungal infections	6 (1.5)	15 (0.4)	4.10 (1.57–10.71)	3.81 (1.45–9.98)
Eye and ear infections	55 (13.9)	215 (5.4)	2.94 (2.12–4.08)	2.93 (2.11–4.07)
Pneumonia	37 (9.3)	150 (3.8)	2.66 (1.82–3.89)	2.69 (1.83–3.95)
URT infections	47 (11.9)	265 (6.7)	1.94 (1.38–2.72)	1.91 (1.36–2.69)
Teeth and oral infections	10 (2.5)	29 (0.7)	3.49 (1.69–7.19)	3.30 (1.59–6.82)
Intraabdominal infections	14 (3.5)	109 (2.8)	1.31 (0.73–2.34)	1.29 (0.72–2.31)
Urinary tract infections	14 (3.5)	85 (2.1)	1.71 (0.95–3.08)	1.69 (0.93–3.04)
Hyperthermia	23 (5.8)	86 (2.2)	2.82 (1.75–4.55)	2.78 (1.72–4.48)
Febrile convulsions	15 (3.8)	86 (2.2)	1.79 (1.02–3.15)	1.79 (1.02–3.15)
Hypohidrosis (n=135)				
Any infection	60 (44.4)	348 (25.8)	2.45 (1.68–3.58)	2.41 (1.65–3.53)
GI tract infections	8 (5.9)	53 (3.9)	1.55 (0.72–3.33)	1.51 (0.70–3.27)
Abscesses	7 (5.2)	42 (3.1)	1.75 (0.75–4.07)	1.78 (0.76–4.17)
Septicaemia	4 (3.0)	4 (0.3)	10.00 (2.50–39.99)	12.03 (2.95–49.14)
Skin and SC infections	10 (7.4)	48 (3.6)	2.29 (1.09–4.78)	2.26 (1.08–4.74)
Viral infections	21 (15.6)	51 (3.8)	5.00 (2.84–8.82)	4.85 (2.75–8.55)
Fungal infections	<3	N/A	N/A	N/A
Eye and ear infections	13 (9.6)	61 (4.5)	2.27 (1.21–4.29)	2.27 (1.21–4.29)
Pneumonia	19 (14.1)	41 (3.0)	5.51 (3.03–10.02)	5.37 (2.95–9.80)
URT infections	20 (14.8)	74 (5.5)	3.04 (1.78–5.18)	3.01 (1.76–5.16)
Teeth and oral infections	4 (3.0)	8 (0.6)	5.00 (1.51–16.60)	4.55 (1.36–15.22)
Intraabdominal infections	5 (3.7)	43 (3.2)	1.18 (0.45–3.13)	1.15 (0.43–3.06)

Urinary tract infections	4 (3.0)	21 (1.6)	1.93 (0.65–5.71)	1.80 (0.61–5.33)
Hyperthermia	11 (8.1)	16 (1.2)	7.11 (3.26–15.51)	7.19 (3.27–15.78)
Febrile convulsions	<3	N/A	N/A	N/A
Incontinencia pigmenti (n=75)				
Any infection	30 (40.0)	159 (21.2)	3.40 (1.86–6.22)	3.58 (1.93–6.61)
GI tract infections	4 (5.3)	12 (1.6)	3.42 (1.08–10.80)	3.49 (1.09–11.19)
Abscesses	<3	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A
Skin and SC infections	10 (13.3)	26 (3.5)	4.44 (2.00–9.82)	4.52 (2.01–10.17)
Viral infections	4 (5.3)	22 (2.9)	1.97 (0.62–6.25)	2.16 (0.67–6.96)
Fungal infections	<3	N/A	N/A	N/A
Eye and ear infections	4 (5.3)	26 (3.5)	1.65 (0.52–5.23)	1.84 (0.57–5.90)
Pneumonia	<3	N/A	N/A	N/A
URT infections	<3	N/A	N/A	N/A
Teeth and oral infections	0	N/A	N/A	N/A
Intraabdominal infections	3 (4.0)	12 (1.6)	2.65 (0.71–9.97)	2.65 (0.70–10.02)
Urinary tract infections	3 (4.0)	17 (2.3)	1.92 (0.50–7.34)	1.91 (0.50–7.30)
Hyperthermia	<3	N/A	N/A	N/A
Febrile convulsions	3 (4.0)	9 (1.2)	3.94 (0.92–16.95)	3.96 (0.92–16.95)
XLHED, males (n=50)				
Any infection	22 (44.0)	114 (22.8)	3.02 (1.57–5.78)	2.96 (1.53–5.71)
GI tract infections	<3	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A
Skin and SC infections	<3	N/A	N/A	N/A
Viral infections	7 (14.0)	22 (4.4)	3.76 (1.46–9.67)	3.63 (1.41–9.38)
Fungal infections	0	N/A	N/A	N/A
Eye and ear infections	5 (10.0)	22 (4.4)	2.50 (0.88–7.14)	2.58 (0.88–7.54)
Pneumonia	9 (18.0)	15 (3.0)	7.30 (2.93–18.21)	7.30 (2.85–18.71)
URT infections	8 (16.0)	33 (6.6)	2.80 (1.18–6.64)	2.79 (1.17–6.64)
Teeth and oral infections	0	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A
Urinary tract infections	0	N/A	N/A	N/A

Hyperthermia	3 (6.0)	5 (1.0)	6.00 (1.43–25.11)	6.03 (1.42–25.68)
Febrile convulsions	0	N/A	N/A	N/A
XLHED, females (n=50)				
Any infection	23 (46.0)	179 (35.8)	1.59 (0.86–2.94)	1.71 (0.91–3.22)
GI tract infections	<3	N/A	N/A	N/A
Abscesses	4 (8.0)	19 (3.8)	2.19 (0.72–6.67)	2.33 (0.74–7.38)
Septicaemia	<3	N/A	N/A	N/A
Skin and SC infections	4 (8.0)	21 (4.2)	2.03 (0.65–6.31)	2.24 (0.69–7.24)
Viral infections	3 (6.0)	19 (3.8)	1.62 (0.46–5.71)	1.52 (0.42–5.45)
Fungal infections	0	N/A	N/A	N/A
Eye and ear infections	6 (12.0)	25 (5.0)	2.60 (1.01–6.73)	2.53 (0.97–6.58)
Pneumonia	4 (8.0)	19 (3.8)	2.19 (0.72–6.67)	3.29 (1.03–10.51)
URT infections	7 (14.0)	32 (6.4)	2.66 (1.03–6.90)	2.55 (0.96–6.78)
Teeth and oral infections	<3	N/A	N/A	N/A
Intraabdominal infections	4 (8.0)	21 (4.2)	2.04 (0.65–6.40)	2.35 (0.72–7.61)
Urinary tract infections	0	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A
Febrile convulsions	<3	N/A	N/A	N/A
Hypohidrotic ED, no genetic diagnosis (n=55)				
Any infection	25 (45.5)	150 (27.3)	2.25 (1.28–3.97)	2.18 (1.23–3.86)
GI tract infections	7 (12.7)	14 (2.5)	5.54 (2.12–14.44)	5.43 (2.06–14.31)
Abscesses	<3	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A
Skin and SC infections	3 (5.5)	17 (3.1)	1.88 (0.51–7.03)	1.77 (0.47–6.66)
Viral infections	10 (18.2)	24 (4.4)	4.94 (2.20–11.08)	4.87 (2.14–11.09)
Fungal infections	0	N/A	N/A	N/A
Eye and ear infections	7 (12.7)	32 (5.8)	2.42 (1.00–5.90)	2.37 (0.97–5.82)
Pneumonia	9 (16.4)	22 (4.0)	4.93 (2.09–11.63)	4.68 (1.97–11.12)
URT infections	8 (14.5)	28 (5.1)	3.15 (1.36–7.28)	2.99 (1.28–7.00)
Teeth and oral infections	3 (5.5)	5 (0.9)	6.00 (1.43–25.11)	5.53 (1.31–23.42)
Intraabdominal infections	<3	N/A	N/A	N/A
Urinary tract infections	<3	N/A	N/A	N/A
Hyperthermia	3 (5.5)	9 (1.6)	3.33 (0.90–12.31)	3.32 (0.89–12.42)

Febrile convulsions	<3	N/A	N/A	N/A
Hidrotic ED, no genetic diagnosis (n=55)				
Any infection	28 (50.9)	214 (38.9)	1.64 (0.94–2.89)	1.67 (0.95–2.96)
GI tract infections	8 (14.5)	34 (6.2)	2.63 (1.14–6.07)	2.52 (1.10–5.77)
Abscesses	<3	N/A	N/A	N/A
Septicaemia	<3	N/A	N/A	N/A
Skin and SC infections	4 (7.3)	34 (6.2)	1.19 (0.40–3.52)	1.22 (0.41–3.61)
Viral infections	10 (18.2)	44 (8.0)	2.64 (1.22–5.71)	2.74 (1.25–6.03)
Fungal infections	<3	N/A	N/A	N/A
Eye and ear infections	9 (16.4)	48 (8.7)	2.06 (0.94–4.49)	1.99 (0.91–4.36)
Pneumonia	4 (7.3)	33 (6.0)	1.23 (0.42–3.61)	1.26 (0.43–3.71)
URT infections	8 (14.5)	53 (9.6)	1.61 (0.72–3.62)	1.60 (0.71–3.62)
Teeth and oral infections	<3	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A
Urinary tract infections	0	N/A	N/A	N/A
Hyperthermia	5 (9.1)	26 (4.7)	2.02 (0.74–5.52)	1.93 (0.71–5.27)
Febrile convulsions	6 (10.9)	25 (4.5)	2.58 (1.01–6.60)	2.48 (0.97–6.37)

^a The number of subgroup prescriptions do not add up to the total, as people may have had multiple categories of prescriptions.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Reference group for OR calculations are the numbers of cases and comparators unexposed of the prescription of interest.

^d Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; GI, gastrointestinal; OR, odds ratio; SC, subcutaneous; URT, upper respiratory tract; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SV. First-time infections and hyperthermia after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events ^a	Person-years at risk	IR per 100,000 person-years	Events	Person-years at risk	IR per 100,000 person-years	Unadjusted ^b	Adjusted ^c
Any ED (n=396)								
Any infection	165	2937	5619	1037	38283	2709	2.06 (1.74–2.45)	2.07 (1.75–2.46)
GI tract infections	18	4494	401	114	45486	251	1.64 (0.99–2.70)	1.64 (0.99–2.71)
Abscesses	18	4465	403	138	45490	303	1.35 (0.82–2.21)	1.21 (0.73–2.00)
Septicaemia	6	4592	131	19	46350	41	3.03 (1.20–7.67)	3.13 (1.22–8.00)
Skin and SC infections	36	4241	849	185	45226	409	2.00 (1.39–2.87)	2.03 (1.41–2.92)
Viral infections	43	4273	1006	186	45157	412	2.41 (1.72–3.38)	2.48 (1.77–3.48)
Fungal infections	12	4514	266	22	46340	47	5.71 (2.79–11.69)	5.39 (2.56–11.33)
Eye and ear infections	39	4211	926	152	45195	336	2.83 (1.98–4.06)	2.91 (2.02–4.17)
Pneumonia	34	4251	800	113	45644	248	3.36 (2.27–4.98)	3.46 (2.32–5.14)
URT infections	33	4364	756	255	44552	572	1.35 (0.93–1.94)	1.37 (0.95–1.97)
Teeth and oral infections	30	4326	693	25	46299	54	13.87 (7.94–24.23)	16.68 (8.80–31.60)
Intraabdominal infections	15	4501	333	108	45664	237	1.36 (0.78–2.35)	1.35 (0.78–2.34)
Urinary tract infections	20	4455	449	127	45777	277	1.60 (0.99–2.58)	1.59 (0.98–2.56)
Hyperthermia	14	4522	310	91	45848	198	1.55 (0.88–2.72)	1.67 (0.94–2.95)
Febrile convulsions	5	4541	110	24	46132	52	2.06 (0.79–5.40)	2.24 (0.84–5.96)

Hypohidrosis (n=135)								
Any infection	70	1089	6426	400	15189	2633	2.29 (1.75–2.99)	2.28 (1.75–2.98)
GI tract infections	6	1778	338	42	18303	229	1.58 (0.66–3.75)	1.57 (0.65–3.79)
Abscesses	8	1762	454	66	18145	364	1.35 (0.64–2.83)	1.21 (0.57–2.57)
Septicaemia	3	1816	165	11	18579	59	2.61 (0.73–9.37)	3.10 (0.78–12.31)
Skin and SC infections	15	1686	890	82	18023	455	1.86 (1.07–3.26)	1.94 (1.10–3.39)
Viral infections	16	1661	963	67	18137	369	2.44 (1.40–4.23)	2.49 (1.42–4.34)
Fungal infections	6	1764	340	6	18621	32	11.51 (3.51–37.79)	14.49 (3.37–62.28)
Eye and ear infections	16	1606	997	61	18083	337	2.90 (1.65–5.09)	2.92 (1.66–5.14)
Pneumonia	17	1642	1036	55	18192	302	3.32 (1.91–5.79)	3.43 (1.92–6.10)
URT infections	13	1690	769	88	17783	495	1.57 (0.87–2.82)	1.50 (0.83–2.71)
Teeth and oral infections	15	1707	879	9	18576	48	24.63 (9.56–63.50)	48.90 (12.09–197.79)
Intraabdominal infections	6	1737	345	57	18203	313	1.12 (0.48–2.62)	1.13 (0.48–2.64)
Urinary tract infections	4	1799	222	38	18466	206	1.00 (0.36–2.81)	1.01 (0.35–2.87)
Hyperthermia	5	1795	279	36	18308	197	1.38 (0.54–3.53)	1.57 (0.60–4.13)
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Incontinentia pigmenti (n=75)								
Any infection	37	562	6581	221	7346	3008	2.40 (1.66–3.46)	2.47 (1.70–3.57)
GI tract infections	6	886	678	31	8845	351	1.98 (0.82–4.77)	2.05 (0.84–4.98)
Abscesses	6	857	700	18	9079	198	3.62 (1.42–9.26)	3.57 (1.35–9.46)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Skin and SC infections	8	827	967	35	8967	390	2.59 (1.19–5.68)	2.94 (1.29–6.71)
Viral infections	13	835	1557	53	8750	606	2.74 (1.47–5.08)	2.95 (1.57–5.55)
Fungal infections	3	909	330	6	9087	66	5.75 (1.37–24.10)	8.96 (1.79–44.99)
Eye and ear infections	8	871	919	37	8869	417	2.63 (1.20–5.77)	2.87 (1.29–6.41)
Pneumonia	4	866	462	24	8941	268	1.78 (0.61–5.17)	1.89 (0.65–5.55)
URT infections	8	880	909	88	8648	1018	0.92 (0.44–1.91)	0.91 (0.44–1.90)
Teeth and oral infections	4	881	454	7	9119	77	6.31 (1.78–22.39)	6.19 (1.63–23.60)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	8	830	964	24	8983	267	3.97 (1.74–9.09)	3.89 (1.66–9.14)
Hyperthermia	5	881	567	24	9024	266	2.11 (0.80–5.54)	2.77 (1.00–7.67)
Febrile convulsions	5	863	579	8	9039	89	6.25 (2.04–19.10)	6.88 (2.02–23.42)
XLHED, males (n=50)								
Any infection	32	398	8039	171	6163	2775	2.60 (1.74–3.88)	2.60 (1.74–3.89)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	7	760	921	45	7463	603	1.40 (0.63–3.10)	1.39 (0.62–3.12)
Viral infections	8	722	1108	35	7549	464	2.20 (1.02–4.78)	2.15 (0.98–4.68)
Fungal infections	N/A	N/A	N/A	<3	N/A	N/A	N/A	N/A
Eye and ear infections	8	706	1133	33	7608	434	2.34 (1.08–5.11)	2.35 (1.07–5.19)
Pneumonia	12	674	1780	17	7676	221	7.66 (3.58–16.36)	7.66 (3.45–16.98)

URT infections	6	742	809	45	7391	609	1.37 (0.58–3.24)	1.37 (0.58–3.24)
Teeth and oral infections	7	756	926	4	7842	51	34.21 (7.10–164.71)	15.38 (4.45–53.17)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
XLHED, females (n=50)								
Any infection	18	398	4519	136	4641	2930	1.58 (0.95–2.63)	1.57 (0.93–2.64)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Viral infections	3	540	555	19	5494	346	1.21 (0.34–4.28)	1.19 (0.32–4.41)
Fungal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Pneumonia	6	498	1205	18	5497	327	4.22 (1.60–11.12)	3.99 (1.35–11.82)
URT infections	4	543	737	19	5445	349	2.03 (0.68–6.03)	2.11 (0.66–6.76)
Teeth and oral infections	3	542	554	4	5552	72	7.12 (1.59–31.91)	7.57 (1.14–50.17)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	4	516	775	20	5469	366	2.11 (0.70–6.35)	2.44 (0.73–8.18)
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Febrile convulsions	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hypohidrotic ED, no genetic diagnosis (n=55)								
Any infection	25	525	4761	143	6744	2120	2.24 (1.44–3.48)	2.07 (1.32–3.25)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	3	688	436	31	7697	403	1.18 (0.35–3.91)	1.05 (0.31–3.57)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	4	705	567	29	7767	373	1.68 (0.58–4.88)	1.66 (0.57–4.88)
Viral infections	5	706	709	18	7844	229	3.03 (1.10–8.35)	3.21 (1.10–9.41)
Fungal infections	N/A	<3	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	5	677	739	22	7718	285	2.65 (0.98–7.15)	2.42 (0.88–6.65)
Pneumonia	3	715	420	22	7829	281	1.47 (0.43–4.99)	1.52 (0.42–5.51)
URT infections	3	694	432	27	7665	352	1.37 (0.41–4.58)	1.46 (0.43–4.99)
Teeth and oral infections	6	703	854	3	7901	38	29.62 (5.98–146.76)	50.19 (5.89–427.89)
Intraabdominal infections	3	698	430	20	7777	257	1.80 (0.52–6.18)	1.80 (0.51–6.34)
Urinary tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hidrotic ED, no genetic diagnosis (n=55)								
Any infection	13	434	2995	124	4681	2649	1.09 (0.61–1.95)	1.09 (0.61–1.95)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	3	489	613	18	5391	334	1.78 (0.52–6.10)	1.58 (0.45–5.49)
Viral infections	4	504	793	18	5350	336	2.62 (0.87–7.90)	2.55 (0.83–7.79)
Fungal infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	3	508	591	15	5353	280	2.36 (0.66–8.35)	2.31 (0.65–8.25)
Pneumonia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
URT infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Teeth and oral infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	3	531	565	15	5456	275	2.37 (0.67–8.42)	2.33 (0.65–8.33)

^a The number of events for individual infections do not add up to the totals, as people may have multiple infections.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; GI, gastrointestinal; HR, hazard ratio; IR, incidence rate; SC, subcutaneous; URT, upper respiratory tract; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SVI. Any infection (both hospital-treated infections and prescriptions) before ectodermal dysplasia diagnosis

Subgroup	ED patients exposed, n (%)	Comparators exposed, n (%)	Unadjusted^a OR^b (95% CI)	Adjusted^c OR^b (95% CI)
Any ED (n=396)	326 (82.3)	2849 (71.9)	3.74 (2.46–5.69)	3.69 (2.42–5.63)
Hypohidrosis (n=135)	114 (84.4)	946 (70.1)	4.63 (2.33–9.19)	4.47 (2.25–8.89)
Incontinentia pigmenti (n=75)	44 (58.7)	346 (46.1)	5.01 (2.07–12.15)	5.14 (2.10–12.57)
XLHED, males (n=50)	37 (74.0)	283 (56.6)	4.08 (1.60–10.40)	4.16 (1.61–10.75)
XLHED, females (n=50)	45 (90.0)	426 (85.2)	2.29 (0.60–8.79)	2.31 (0.59–9.09)
Hypohidrotic ED, no genetic diagnosis (n=55)	52 (94.5)	425 (77.3)	7.64 (2.13–27.41)	7.25 (2.00–26.28)
Hidrotic ED, no genetic diagnosis (n=55)	49 (89.1)	493 (89.6)	0.94 (0.38–2.34)	0.83 (0.33–2.09)

^a Adjusted for matching factors (age, sex, and municipality) by design.

^b Reference group for OR calculations are the numbers of cases and comparators unexposed of the prescription of interest.

^c Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; OR, odds ratio; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SVII. Any first-time infection (both hospital-treated infections and prescriptions) after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events	Person-years at risk	IR per 100,000 person-years	Events	Person-years at risk	IR per 100,000 person-years	Unadjusted ^a	Adjusted ^b
Any ED (n=396)	312	874	35688	3045	12728	23924	1.43 (1.26–1.62)	1.42 (1.26–1.62)
Hypohidrosis (n=135)	116	252	45991	1131	4780	23663	1.75 (1.42–2.16)	1.74 (1.41–2.15)
Incontinentia pigmenti (n=75)	61	145	41980	586	2039	28736	1.36 (1.02–1.80)	1.39 (1.05–1.86)
XLHED, males (n=50)	43	87	49203	429	1685	25462	2.10 (1.48–3.00)	2.13 (1.49–3.05)
XLHED, females (n=50)	43	109	39398	421	1502	28031	1.35 (0.96–1.90)	1.35 (0.96–1.90)
Hypohidrotic ED, no genetic diagnosis (n=55)	49	114	43044	469	2226	21073	1.73 (1.25–2.39)	1.72 (1.24–2.38)
Hidrotic ED, no genetic diagnosis (n=55)	40	164	24353	384	1878	20452	1.20 (0.84–1.71)	1.19 (0.84–1.70)

^a Adjusted for matching factors (age, sex, and municipality) by design.

^b Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; HR, hazard ratio; IR, incidence rate; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SVIII. Prescriptions of antimicrobials before ectodermal dysplasia diagnosis

Subgroup	ED patients exposed ^a , n (%)	Comparators exposed, n (%)	Unadjusted ^b OR ^c (95% CI)	Adjusted ^d OR ^c (95% CI)
Any ED (n=396)				
Systemic antibiotics, any	297 (75.0)	2696 (68.1)	2.28 (1.53–3.38)	2.22 (1.50–3.30)
Phenoxymethylpenicillin	247 (62.4)	2205 (55.7)	1.60 (1.20–2.12)	1.56 (1.17–2.08)
Aminopenicillins	175 (44.2)	1397 (35.3)	1.70 (1.32–2.18)	1.69 (1.31–2.17)
Macrolides	141 (35.6)	1065 (26.9)	1.70 (1.32–2.19)	1.67 (1.30–2.16)
Cystitis antibiotics	86 (21.7)	656 (16.6)	1.68 (1.22–2.33)	1.70 (1.23–2.35)
BL resistant penicillins	80 (20.2)	471 (11.9)	2.06 (1.55–2.75)	2.06 (1.54–2.74)
Quinolones	25 (6.3)	159 (4.0)	1.70 (1.07–2.70)	1.63 (1.03–2.60)
Systemic antimycotics	46 (11.6)	387 (9.8)	1.30 (0.89–1.91)	1.28 (0.87–1.89)
Systemic antivirals	25 (6.3)	185 (4.7)	1.40 (0.90–2.18)	1.37 (0.88–2.14)
Ophthalmic antimicrobials	221 (55.8)	1919 (48.5)	1.57 (1.21–2.04)	1.54 (1.19–2.00)
Otological antimicrobials	91 (23.0)	443 (11.2)	2.64 (2.00–3.47)	2.60 (1.97–3.42)
Hypohidrosis (n=135)				
Systemic antibiotics, any	106 (78.5)	889 (65.9)	3.52 (1.84–6.75)	3.44 (1.80–6.57)
Phenoxymethylpenicillin	84 (62.2)	702 (52.0)	1.92 (1.20–3.05)	1.89 (1.19–3.01)
Aminopenicillins	58 (43.0)	410 (30.4)	2.09 (1.37–3.20)	2.02 (1.32–3.11)
Macrolides	48 (35.6)	345 (25.6)	1.82 (1.18–2.79)	1.81 (1.18–2.78)
Cystitis antibiotics	25 (18.5)	176 (13.0)	1.92 (1.06–3.49)	1.97 (1.08–3.59)
BL resistant penicillins	18 (13.3)	139 (10.3)	1.40 (0.79–2.47)	1.42 (0.80–2.50)
Quinolones	10 (7.4)	46 (3.4)	2.45 (1.15–5.22)	2.53 (1.18–5.42)
Systemic antimycotics	14 (10.4)	116 (8.6)	1.32 (0.67–2.61)	1.34 (0.68–2.66)
Systemic antivirals	10 (7.4)	54 (4.0)	1.97 (0.96–4.04)	2.00 (0.97–4.11)
Ophthalmic antimicrobials	68 (50.4)	590 (43.7)	1.44 (0.95–2.19)	1.42 (0.93–2.15)
Otological antimicrobials	22 (16.3)	122 (9.0)	2.11 (1.25–3.56)	2.03 (1.20–3.43)
Incontinentia pigmenti (n=75)				
Systemic antibiotics, any	38 (50.7)	315 (42.0)	5.39 (1.56–18.63)	5.52 (1.55–19.61)
Phenoxymethylpenicillin	30 (40.0)	255 (34.0)	1.99 (0.85–4.68)	1.85 (0.77–4.42)
Aminopenicillins	19 (25.3)	134 (17.9)	1.96 (0.97–3.95)	1.91 (0.94–3.86)

Macrolides	20 (26.7)	152 (20.3)	1.84 (0.88–3.85)	1.79 (0.85–3.79)
Cystitis antibiotics	19 (25.3)	139 (18.5)	1.98 (0.92–4.24)	1.99 (0.92–4.30)
BL resistant penicillins	10 (13.3)	56 (7.5)	2.11 (0.96–4.66)	2.20 (0.98–4.93)
Quinolones	4 (5.3)	35 (4.7)	1.18 (0.37–3.75)	1.03 (0.32–3.32)
Systemic antimycotics	10 (13.3)	96 (12.8)	1.06 (0.47–2.39)	0.99 (0.44–2.23)
Systemic antivirals	6 (8.0)	33 (4.4)	2.03 (0.77–5.33)	1.98 (0.76–5.20)
Ophthalmic antimicrobials	27 (36.0)	212 (28.3)	1.94 (0.95–3.96)	1.88 (0.92–3.85)
Otological antimicrobials	11 (14.7)	38 (5.1)	3.99 (1.76–9.01)	4.14 (1.79–9.56)
XLHED, males (n=50)				
Systemic antibiotics, any	31 (62.0)	257 (51.4)	2.16 (0.93–4.97)	2.15 (0.92–5.01)
Phenoxymethylpenicillin	23 (46.0)	181 (36.2)	1.85 (0.89–3.83)	1.86 (0.89–3.87)
Aminopenicillins	16 (32.0)	128 (25.6)	1.57 (0.73–3.37)	1.57 (0.73–3.38)
Macrolides	18 (36.0)	86 (17.2)	4.30 (1.96–9.45)	4.09 (1.86–8.96)
Cystitis antibiotics	0	N/A	N/A	N/A
BL resistant penicillins	<3	N/A	N/A	N/A
Quinolones	<3	N/A	N/A	N/A
Systemic antimycotics	<3	N/A	N/A	N/A
Systemic antivirals	<3	N/A	N/A	N/A
Ophthalmic antimicrobials	20 (40.0)	179 (35.8)	1.29 (0.63–2.66)	1.27 (0.62–2.62)
Otological antimicrobials	5 (10.0)	24 (4.8)	2.29 (0.80–6.53)	2.18 (0.76–6.25)
XLHED, females (n=50)				
				2.47 (0.58–
Systemic antibiotics, any	44 (88.0)	412 (82.4)	2.69 (0.66–11.00)	10.56)
Phenoxymethylpenicillin	37 (74.0)	339 (67.8)	1.55 (0.69–3.49)	1.36 (0.59–3.09)
Aminopenicillins	27 (54.0)	194 (38.8)	2.12 (1.11–4.05)	2.24 (1.15–4.36)
Macrolides	28 (56.0)	198 (39.6)	2.30 (1.17–4.51)	2.34 (1.17–4.69)
Cystitis antibiotics	17 (34.0)	174 (34.8)	0.96 (0.49–1.88)	0.99 (0.50–1.96)
BL resistant penicillins	13 (26.0)	86 (17.2)	1.77 (0.87–3.59)	1.85 (0.89–3.82)
Quinolones	5 (10.0)	41 (8.2)	1.25 (0.46–3.36)	1.04 (0.38–2.85)
Systemic antimycotics	11 (22.0)	97 (19.4)	1.21 (0.56–2.62)	1.19 (0.55–2.60)
Systemic antivirals	5 (10.0)	34 (6.8)	1.54 (0.56–4.21)	1.65 (0.58–4.66)
Ophthalmic antimicrobials	26 (52.0)	253 (50.6)	1.07 (0.56–2.04)	1.07 (0.56–2.06)
Otological antimicrobials	11 (22.0)	64 (12.8)	2.06 (0.96–4.41)	1.96 (0.92–4.20)

Hypohidrotic ED, no genetic diagnosis (n=55)				
				3.84 (1.35–10.92)
Systemic antibiotics, any	47 (85.5)	396 (72.0)	4.03 (1.41–11.48)	
Phenoxymethylpenicillin	36 (65.5)	322 (58.5)	1.47 (0.75–2.89)	1.40 (0.72–2.76)
Aminopenicillins	27 (49.1)	180 (32.7)	2.64 (1.33–5.23)	2.49 (1.25–4.98)
Macrolides	12 (21.8)	147 (26.7)	0.74 (0.37–1.49)	0.74 (0.37–1.50)
Cystitis antibiotics	11 (20.0)	67 (12.2)	2.38 (0.99–5.75)	2.46 (1.02–5.95)
BL resistant penicillins	9 (16.4)	57 (10.4)	1.79 (0.79–4.06)	1.84 (0.81–4.19)
Quinolones	<3	N/A	N/A	N/A
Systemic antimycotics	5 (9.1)	44 (8.0)	1.23 (0.37–4.03)	1.21 (0.37–3.99)
Systemic antivirals	<3	N/A	N/A	N/A
Ophthalmic antimicrobials	31 (56.4)	260 (47.3)	1.63 (0.85–3.13)	1.55 (0.81–3.00)
Otological antimicrobials	11 (20.0)	61 (11.1)	2.26 (1.04–4.90)	2.08 (0.95–4.52)
Hidrotic ED, no genetic diagnosis (n=55)				
Systemic antibiotics, any	46 (83.6)	475 (86.4)	0.80 (0.36–1.74)	0.73 (0.33–1.61)
Phenoxymethylpenicillin	42 (76.4)	393 (71.5)	1.33 (0.67–2.62)	1.20 (0.61–2.39)
Aminopenicillins	28 (50.9)	276 (50.2)	1.03 (0.57–1.88)	1.03 (0.56–1.89)
Macrolides	17 (30.9)	157 (28.5)	1.14 (0.60–2.17)	1.12 (0.59–2.14)
Cystitis antibiotics	8 (14.5)	62 (11.3)	1.53 (0.58–4.00)	1.52 (0.57–4.08)
BL resistant penicillins	15 (27.3)	74 (13.5)	2.52 (1.30–4.88)	2.48 (1.27–4.82)
Quinolones	3 (5.5)	15 (2.7)	2.14 (0.57–8.00)	1.97 (0.52–7.46)
Systemic antimycotics	4 (7.3)	29 (5.3)	1.63 (0.43–6.09)	1.47 (0.39–5.61)
Systemic antivirals	<3	N/A	N/A	N/A
Ophthalmic antimicrobials	39 (70.9)	369 (67.1)	1.25 (0.63–2.45)	1.13 (0.57–2.24)
Otological antimicrobials	19 (34.5)	92 (16.7)	2.71 (1.47–5.00)	2.63 (1.41–4.90)

^a The number of subgroup prescriptions do not add up to the total, as people may have had multiple categories of prescriptions.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Reference group for OR calculations are the numbers of cases and comparators unexposed of the prescription of interest.

^d Adjusted for educational level.

Abbreviations: BL, beta-lactamase; CI, confidence interval; ED, ectodermal dysplasia; OR, odds ratio; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SIX. First-time prescriptions of antimicrobials after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events ^a	Person-years at risk	IR per 100,000 person years	Events	Person-years at risk	IR per 100,000 person years	Unadjusted ^b	Adjusted ^c
Any ED (n=396)								
Systemic antibiotics, any	313	1028	30458	2944	13731	21440	1.36 (1.20–1.55)	1.36 (1.20–1.54)
Phenoxymethylpenicillin	255	1769	14418	2288	21433	10675	1.38 (1.20–1.58)	1.38 (1.20–1.58)
Aminopenicillins	168	2770	6065	1086	34927	3109	1.93 (1.63–2.29)	1.95 (1.65–2.31)
Macrolides	139	3067	4533	1263	33578	3761	1.21 (1.01–1.45)	1.20 (1.00–1.44)
Cystitis antibiotics	105	3720	2822	961	38610	2489	1.21 (0.98–1.49)	1.20 (0.98–1.48)
BL resistant penicillins	123	3493	3522	836	39659	2108	1.66 (1.37–2.01)	1.63 (1.34–1.97)
Quinolones	44	4205	1046	259	44414	583	1.77 (1.28–2.44)	1.79 (1.29–2.48)
Systemic antimycotics	68	3969	1713	487	42299	1151	1.47 (1.14–1.91)	1.49 (1.15–1.94)
Systemic antivirals	56	4152	1349	326	43893	743	1.71 (1.28–2.27)	1.73 (1.29–2.30)
Ophthalmic antimicrobials	211	2368	8912	1672	28031	5965	1.51 (1.30–1.75)	1.50 (1.29–1.74)
Otological antimicrobials	74	3889	1903	401	42078	953	1.97 (1.53–2.53)	1.97 (1.53–2.53)
Hypohidrosis (n=135)								
Systemic antibiotics, any	115	295	39047	1104	5105	21626	1.65 (1.34–2.04)	1.63 (1.33–2.02)
Phenoxymethylpenicillin	102	575	17741	891	8070	11041	1.68 (1.35–2.10)	1.67 (1.34–2.08)
Aminopenicillins	69	952	7251	426	13830	3080	2.20 (1.68–2.88)	2.17 (1.66–2.84)

Macrolides	67	980	6839	483	13248	3646	1.85 (1.42–2.42)	1.84 (1.40–2.40)
Cystitis antibiotics	26	1558	1668	291	16224	1794	1.00 (0.66–1.50)	1.03 (0.68–1.55)
BL resistant penicillins	55	1309	4202	334	15626	2138	1.95 (1.45–2.61)	1.90 (1.42–2.56)
Quinolones	19	1633	1163	100	17784	562	2.16 (1.31–3.57)	2.15 (1.30–3.55)
Systemic antimycotics	21	1580	1329	146	17312	843	1.65 (1.03–2.63)	1.70 (1.06–2.72)
Systemic antivirals	26	1577	1649	101	17837	566	2.78 (1.79–4.32)	2.85 (1.83–4.44)
Ophthalmic antimicrobials	72	1065	6762	638	10996	5802	1.24 (0.97–1.60)	1.24 (0.97–1.60)
Otological antimicrobials	22	1582	1391	151	16879	895	1.56 (0.99–2.45)	1.52 (0.97–2.40)
Incontinencia pigmenti (n=75)								
Systemic antibiotics, any	62	173	35815	557	2277	24458	1.34 (1.01–1.79)	1.38 (1.03–1.84)
Phenoxymethylpenicillin	46	409	11244	451	3669	12292	0.91 (0.67–1.25)	0.91 (0.66–1.25)
Aminopenicillins	37	494	7496	279	5887	4739	1.62 (1.13–2.31)	1.63 (1.14–2.33)
Macrolides	26	658	3954	243	6541	3715	1.06 (0.70–1.61)	1.03 (0.68–1.56)
Cystitis antibiotics	32	608	5267	236	7057	3344	1.77 (1.21–2.61)	1.78 (1.20–2.62)
BL resistant penicillins	23	703	3274	143	7906	1809	1.94 (1.23–3.05)	1.85 (1.17–2.92)
Quinolones	8	849	943	45	8786	512	1.75 (0.82–3.75)	1.82 (0.84–3.92)
Systemic antimycotics	15	794	1890	101	8199	1232	1.54 (0.88–2.68)	1.77 (0.99–3.15)
Systemic antivirals	9	873	1031	73	8631	846	1.11 (0.55–2.22)	1.07 (0.54–2.15)
Ophthalmic antimicrobials	55	294	18684	385	4439	8672	1.75 (1.29–2.37)	1.74 (1.28–2.36)
Otological antimicrobials	17	749	2270	101	7961	1269	1.66 (0.99–2.79)	1.68 (0.99–2.83)
XLHED, males (n=50)								

Systemic antibiotics, any	44	106	41437	418	1800	23225	1.92 (1.36–2.71)	1.95 (1.37–2.76)
Phenoxymethylpenicillin	43	172	25037	363	2917	12446	2.06 (1.46–2.91)	2.04 (1.44–2.89)
Aminopenicillins	29	410	7077	180	5325	3380	2.21 (1.46–3.34)	2.24 (1.47–3.41)
Macrolides	28	396	7072	189	5424	3485	1.93 (1.27–2.93)	1.90 (1.24–2.89)
Cystitis antibiotics	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
BL resistant penicillins	24	568	4226	133	6458	2059	1.85 (1.18–2.89)	1.74 (1.10–2.74)
Quinolones	4	754	531	35	7571	462	1.26 (0.44–3.58)	1.20 (0.42–3.45)
Systemic antimycotics	4	773	518	27	7667	352	1.53 (0.53–4.41)	1.47 (0.50–4.31)
Systemic antivirals	14	684	2046	26	7670	339	5.61 (2.88–10.92)	6.51 (3.24–13.08)
Ophthalmic antimicrobials	30	417	7189	266	4309	6174	1.28 (0.86–1.89)	1.25 (0.84–1.86)
Otological antimicrobials	12	682	1760	67	7015	955	1.99 (1.07–3.72)	1.98 (1.05–3.73)
XLHED, females (n=50)								
Systemic antibiotics, any	42	152	27658	412	1621	25409	1.18 (0.84–1.65)	1.18 (0.84–1.67)
Phenoxymethylpenicillin	33	210	15689	307	2622	11710	1.26 (0.86–1.83)	1.26 (0.86–1.84)
Aminopenicillins	18	371	4848	128	4508	2839	1.61 (0.97–2.67)	1.58 (0.94–2.65)
Macrolides	19	377	5040	177	4051	4369	1.18 (0.73–1.93)	1.15 (0.70–1.88)
Cystitis antibiotics	18	429	4199	204	3989	5114	0.87 (0.53–1.41)	0.90 (0.55–1.47)
BL resistant penicillins	15	440	3409	121	4744	2550	1.29 (0.74–2.22)	1.22 (0.70–2.13)
Quinolones	9	492	1831	41	5317	771	2.20 (1.05–4.58)	2.67 (1.20–5.95)
Systemic antimycotics	12	449	2671	83	4912	1690	1.47 (0.80–2.72)	1.36 (0.72–2.55)
Systemic antivirals	6	490	1224	48	5248	915	1.28 (0.54–3.01)	1.42 (0.60–3.38)

Ophthalmic antimicrobials	18	396	4543	200	3758	5322	0.90 (0.55–1.48)	0.91 (0.55–1.51)
Otological antimicrobials	4	514	778	40	5255	761	1.19 (0.42–3.38)	1.31 (0.45–3.76)
Hypohidrotic ED, no genetic diagnosis (n=55)								
Systemic antibiotics, any	49	126	39020	458	2406	19035	1.77 (1.28–2.44)	1.76 (1.27–2.42)
Phenoxymethylpenicillin	43	257	16756	355	3708	9574	1.88 (1.34–2.64)	1.88 (1.34–2.63)
Aminopenicillins	24	457	5248	154	6474	2379	2.13 (1.36–3.35)	2.03 (1.28–3.20)
Macrolides	26	453	5736	228	5418	4208	1.38 (0.91–2.10)	1.37 (0.90–2.08)
Cystitis antibiotics	17	523	3253	142	6579	2158	1.76 (1.04–2.97)	1.78 (1.05–3.02)
BL resistant penicillins	21	539	3895	137	6746	2031	2.07 (1.28–3.33)	2.00 (1.24–3.23)
Quinolones	8	637	1256	44	7532	584	2.05 (0.95–4.42)	2.01 (0.93–4.35)
Systemic antimycotics	13	589	2206	89	7052	1262	1.86 (1.02–3.41)	1.95 (1.06–3.58)
Systemic antivirals	9	662	1359	55	7457	738	1.71 (0.84–3.51)	1.86 (0.90–3.86)
Ophthalmic antimicrobials	30	444	6759	238	5125	4644	1.41 (0.95–2.09)	1.39 (0.94–2.07)
Otological antimicrobials	8	641	1248	52	7311	711	1.52 (0.72–3.21)	1.54 (0.73–3.28)
Hidrotic ED, no genetic diagnosis (n=55)								
Systemic antibiotics, any	41	174	23591	368	2013	18283	1.22 (0.86–1.72)	1.20 (0.84–1.70)
Phenoxymethylpenicillin	33	254	13007	278	2973	9352	1.44 (0.98–2.09)	1.43 (0.98–2.09)
Aminopenicillins	17	415	4099	102	4618	2209	2.03 (1.19–3.47)	2.06 (1.20–3.53)
Macrolides	11	422	2608	168	4049	4149	0.64 (0.35–1.20)	0.64 (0.35–1.20)
Cystitis antibiotics	13	445	2919	118	4588	2572	1.11 (0.62–1.99)	1.13 (0.62–2.04)
BL resistant penicillins	9	456	1972	106	4658	2275	0.83 (0.42–1.64)	0.80 (0.40–1.60)

Quinolones	4	506	790	34	5207	653	1.12 (0.39–3.16)	1.06 (0.37–3.04)
Systemic antimycotics	4	499	801	70	4973	1408	0.54 (0.19–1.48)	0.51 (0.18–1.42)
Systemic antivirals	5	494	1012	46	5098	902	1.14 (0.45–2.89)	1.08 (0.42–2.75)
Ophthalmic antimicrobials	20	323	6185	188	3892	4830	1.42 (0.88–2.29)	1.38 (0.85–2.22)
Otological antimicrobials	7	480	1457	33	5139	642	2.12 (0.93–4.83)	2.28 (0.98–5.28)

^a The number of events for individual prescriptions do not add up to the totals, as people may have multiple categories of prescriptions.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Adjusted for educational level.

Abbreviations: BL, beta-lactamase; CI, confidence interval; ED, ectodermal dysplasia; HR, hazard ratio; IR, incidence rate; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SX. Infections and hyperthermia (inpatients only) before ectodermal dysplasia diagnosis

Subgroup	ED patients exposed ^a , n (%)	Comparators exposed, n (%)	Unadjusted ^b OR ^c (95% CI)	Adjusted ^d OR ^c (95% CI)
Any ED (n=396)				
Any infection	154 (38.9)	927 (23.4)	2.23 (1.78–2.80)	2.21 (1.76–2.78)
GI tract infections	29 (7.3)	140 (3.5)	2.19 (1.44–3.33)	2.16 (1.42–3.29)
Abscesses	10 (2.5)	86 (2.2)	1.17 (0.60–2.29)	1.17 (0.60–2.30)
Septicaemia	6 (1.5)	14 (0.4)	4.29 (1.65–11.15)	4.17 (1.60–10.87)
Skin and SC infections	26 (6.6)	96 (2.4)	2.85 (1.82–4.47)	2.90 (1.84–4.55)
Viral infections	41 (10.4)	152 (3.8)	3.07 (2.11–4.47)	3.00 (2.06–4.38)
Fungal infections	3 (0.8)	10 (0.3)	3.00 (0.83–10.90)	2.77 (0.76–10.13)
Eye and ear infections	34 (8.6)	129 (3.3)	2.93 (1.95–4.41)	2.94 (1.95–4.42)
Pneumonia	34 (8.6)	142 (3.6)	2.57 (1.73–3.82)	2.59 (1.74–3.86)
URT infections	32 (8.1)	210 (5.3)	1.59 (1.07–2.35)	1.56 (1.05–2.31)
Teeth and oral infections	4 (1.0)	18 (0.5)	2.22 (0.75–6.57)	2.08 (0.70–6.15)
Intraabdominal infections	12 (3.0)	100 (2.5)	1.22 (0.65–2.26)	1.20 (0.64–2.24)
Urinary tract infections	12 (3.0)	58 (1.5)	2.12 (1.12–4.01)	2.09 (1.10–3.95)
Hyperthermia	18 (4.5)	63 (1.6)	2.98 (1.74–5.12)	2.98 (1.73–5.12)
Febrile convulsions	14 (3.5)	80 (2.0)	1.80 (1.00–3.23)	1.80 (1.00–3.23)
Hypohidrosis (n=135)				
Any infection	55 (40.7)	279 (20.7)	2.79 (1.90–4.09)	2.75 (1.87–4.03)
GI tract infections	7 (5.2)	48 (3.6)	1.48 (0.66–3.33)	1.45 (0.64–3.28)
Abscesses	5 (3.7)	33 (2.4)	1.54 (0.59–4.01)	1.57 (0.60–4.11)
Septicaemia	4 (3.0)	4 (0.3)	10.00 (2.50–39.99)	12.03 (2.95–49.14)
Skin and SC infections	8 (5.9)	28 (2.1)	3.00 (1.33–6.76)	3.00 (1.32–6.78)
Viral infections	20 (14.8)	35 (2.6)	6.79 (3.73–12.33)	6.58 (3.60–12.01)
Fungal infections	<3	N/A	N/A	N/A
Eye and ear infections	8 (5.9)	33 (2.4)	2.62 (1.15–5.94)	2.62 (1.15–5.94)
Pneumonia	18 (13.3)	40 (3.0)	5.36 (2.90–9.88)	5.23 (2.83–9.67)
URT infections	17 (12.6)	63 (4.7)	2.95 (1.67–5.21)	2.94 (1.65–5.21)
Teeth and oral infections	<3	N/A	N/A	N/A
Intraabdominal infections	5 (3.7)	41 (3.0)	1.24 (0.47–3.30)	1.22 (0.46–3.25)

Urinary tract infections	3 (2.2)	16 (1.2)	1.89 (0.55–6.56)	1.69 (0.49–5.89)
Hyperthermia	9 (6.7)	11 (0.8)	8.18 (3.39–19.74)	8.37 (3.45–20.34)
Febrile convulsions	<3	N/A	N/A	N/A
Incontinencia pigmenti (n=75)				
Any infection	25 (33.3)	120 (16.0)	3.24 (1.79–5.87)	3.39 (1.85–6.22)
GI tract infections	4 (5.3)	11 (1.5)	3.75 (1.17–12.03)	3.84 (1.17–12.54)
Abscesses	<3	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A
Skin and SC infections	8 (10.7)	13 (1.7)	7.00 (2.73–17.91)	7.63 (2.90–20.06)
Viral infections	3 (4.0)	17 (2.3)	1.94 (0.50–7.48)	2.06 (0.53–8.05)
Fungal infections	<3	N/A	N/A	N/A
Eye and ear infections	0	N/A	N/A	N/A
Pneumonia	<3	N/A	N/A	N/A
URT infections	<3	N/A	N/A	N/A
Teeth and oral infections	0	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A
Urinary tract infections	<3	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A
Febrile convulsions	3 (4.0)	9 (1.2)	3.94 (0.92–16.95)	3.96 (0.92–16.95)
XLHED, males (n=50)				
Any infection	22 (44.0)	93 (18.6)	4.03 (2.08–7.81)	3.94 (2.01–7.71)
GI tract infections	<3	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A
Skin and SC infections	<3	N/A	N/A	N/A
Viral infections	7 (14.0)	14 (2.8)	5.87 (2.20–15.71)	5.89 (2.17–15.98)
Fungal infections	0	N/A	N/A	N/A
Eye and ear infections	3 (6.0)	10 (2.0)	3.37 (0.84–13.46)	3.58 (0.86–14.79)
Pneumonia	9 (18.0)	14 (2.8)	8.04 (3.15–20.55)	7.99 (3.05–20.94)
URT infections	7 (14.0)	28 (5.6)	2.84 (1.14–7.05)	2.79 (1.12–6.97)
Teeth and oral infections	0	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A
Urinary tract infections	0	N/A	N/A	N/A

Hyperthermia	<3	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A

^a The number of subgroup prescriptions do not add up to the total, as people may have had multiple categories of infections.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^b Reference group for OR calculations are the numbers of cases and comparators unexposed of the infection of interest.

^c Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; GI, gastrointestinal; OR, odds ratio; SC, subcutaneous; URT, upper respiratory tract; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SXI. First-time infections and hyperthermia (inpatients only) after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events ^a	Person-years at risk	IR per 100,000 person-years	Events	Person-years at risk	IR per 100,000 person-years	Unadjusted ^b	Adjusted ^c
Any ED (n=396)								
Any infection	92	3640	2527	556	41285	1347	1.92 (1.53–2.40)	1.91 (1.52–2.40)
GI tract infections	13	4510	288	77	45661	169	1.69 (0.94–3.04)	1.69 (0.94–3.06)
Abscesses	11	4544	242	75	45847	164	1.47 (0.78–2.79)	1.35 (0.71–2.58)
Septicaemia	6	4592	131	16	46369	35	3.68 (1.42–9.53)	3.73 (1.43–9.74)
Skin and SC infections	12	4502	267	67	45957	146	1.73 (0.93–3.21)	1.83 (0.97–3.44)
Viral infections	22	4412	499	80	45722	175	2.83 (1.75–4.59)	3.03 (1.86–4.95)
Fungal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	15	4442	338	35	46036	76	4.52 (2.45–8.36)	5.18 (2.75–9.75)
Pneumonia	27	4317	625	78	45800	170	3.77 (2.41–5.90)	3.75 (2.38–5.92)
URT infections	19	4439	428	103	45386	227	1.94 (1.18–3.17)	1.93 (1.17–3.16)
Teeth and oral infections	8	4535	176	5	46412	11	19.11 (5.75–63.54)	64.34 (7.40–559.48)
Intraabdominal infections	14	4505	311	99	45712	217	1.39 (0.79–2.47)	1.39 (0.79–2.47)
Urinary tract infections	13	4494	289	65	46065	141	2.04 (1.12–3.73)	2.00 (1.09–3.67)
Hyperthermia	6	4563	132	52	46052	113	1.14 (0.49–2.66)	1.37 (0.58–3.26)
Febrile convulsions	3	4543	66	23	46147	50	1.29 (0.39–4.29)	1.43 (0.42–4.82)

Hypohidrosis (n=135)								
Any infection	40	1389	2880	216	16469	1312	2.17 (1.54–3.07)	2.16 (1.52–3.07)
GI tract infections	6	1778	338	24	18399	130	2.52 (1.03–6.20)	2.45 (0.95–6.34)
Abscesses	4	1807	221	37	18341	202	1.16 (0.41–3.30)	1.05 (0.37–2.99)
Septicaemia	3	1816	165	8	18599	43	3.71 (0.99–14.00)	4.25 (0.96–18.89)
Skin and SC infections	5	1782	281	31	18418	168	1.62 (0.63–4.19)	1.64 (0.62–4.29)
Viral infections	13	1704	763	25	18427	136	5.63 (2.83–11.17)	5.86 (2.89–11.87)
Fungal infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	7	1731	404	17	18419	92	4.19 (1.72–10.18)	4.62 (1.84–11.60)
Pneumonia	13	1679	774	38	18276	208	3.53 (1.86–6.70)	3.35 (1.68–6.66)
URT infections	9	1730	520	37	18162	204	2.78 (1.32–5.84)	2.73 (1.27–5.86)
Teeth and oral infections	N/A	N/A	N/A	<3	N/A	N/A	N/A	N/A
Intraabdominal infections	6	1737	345	51	18247	280	1.29 (0.55–3.03)	1.31 (0.56–3.09)
Urinary tract infections	4	1799	222	20	18549	108	1.92 (0.65–5.65)	1.60 (0.53–4.86)
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Incontinentia pigmenti (n=75)								
Any infection	23	666	3451	128	7887	1623	2.19 (1.39–3.47)	2.25 (1.42–3.57)
GI tract infections	4	893	448	23	8873	259	1.75 (0.60–5.08)	1.81 (0.62–5.32)
Abscesses	3	889	337	10	9102	110	2.87 (0.79–10.45)	3.93 (0.97–15.98)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Skin and SC infections	3	892	336	9	9076	99	3.21 (0.87–11.87)	4.81 (1.13–20.45)
Viral infections	5	868	576	25	8881	282	2.16 (0.82–5.70)	2.61 (0.95–7.17)
Fungal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Pneumonia	4	866	462	18	8957	201	2.30 (0.78–6.85)	2.32 (0.78–6.93)
URT infections	3	900	333	33	8902	371	0.92 (0.28–3.02)	0.92 (0.28–3.01)
Teeth and oral infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	6	837	717	14	9056	155	5.35 (1.98–14.48)	6.73 (2.22–20.39)
Hyperthermia	3	886	338	13	9059	144	2.29 (0.65–8.04)	4.95 (1.13–21.71)
Febrile convulsions	3	865	347	7	9054	77	4.29 (1.11–16.57)	5.39 (1.25–23.23)
XLHED, males (n=50)								
Any infection	15	610	2461	88	6826	1289	1.92 (1.10–3.37)	1.86 (1.05–3.29)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Viral infections	5	763	655	11	7767	142	4.74 (1.62–13.87)	4.80 (1.54–14.93)
Fungal infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Pneumonia	8	711	1125	13	7692	169	6.25 (2.55–15.32)	5.17 (1.99–13.40)

URT infections	4	764	523	18	7562	238	2.40 (0.80–7.18)	3.04 (0.95–9.77)
Teeth and oral infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A

^a The number of events for individual infections do not add up to the totals, as people may have multiple infections.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; GI, gastrointestinal; HR, hazard ratio; IR, incidence rate; SC, subcutaneous; URT, upper respiratory tract; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SXII. Recurrent infections and hyperthermia after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events ^a	Person-years at risk	IR per 100,000 person-years	Events	Person-years at risk	IR per 100,000 person-years	Unadjusted ^b	Adjusted ^c
Any ED (n=396)								
Any infection	596	4665	12775	1933	46623	4146	3.04 (2.53–3.65)	3.05 (2.55–3.65)
GI tract infections	32	4627	692	133	46459	286	2.43 (1.51–3.91)	2.44 (1.50–3.97)
Abscesses	29	4627	627	201	46484	432	1.44 (0.82–2.54)	1.30 (0.73–2.33)
Septicaemia	14	4627	303	23	46459	50	5.21 (2.01–13.48)	5.16 (2.06–12.90)
Skin and SC infections	68	4629	1469	244	46472	525	2.72 (1.86–3.97)	2.76 (1.88–4.06)
Viral infections	67	4646	1442	234	46493	503	2.76 (1.95–3.89)	2.79 (1.99–3.92)
Fungal infections	14	4627	303	23	46459	50	6.33 (3.30–12.15)	6.37 (3.19–12.70)
Eye and ear infections	68	4629	1469	244	46472	525	2.72 (1.86–3.97)	2.76 (1.88–4.06)
Pneumonia	84	4638	1811	142	46470	306	5.73 (4.02–8.17)	5.90 (4.11–8.48)
URT infections	58	4643	1249	328	46477	706	1.79 (1.30–2.47)	1.82 (1.33–2.50)
Teeth and oral infections	43	4631	929	28	46471	60	16.09 (10.34–25.02)	19.21 (11.41–32.33)
Intraabdominal infections	19	4639	410	126	46470	271	1.44 (0.86–2.42)	1.44 (0.88–2.36)
Urinary tract infections	38	4627	821	181	46477	389	2.02 (1.28–3.19)	1.99 (1.27–3.12)
Hyperthermia	22	4627	476	105	46485	226	2.06 (1.06–4.00)	2.23 (1.22–4.07)
Febrile convulsions	9	4627	195	33	46459	71	2.71 (1.13–6.47)	2.79 (1.14–6.81)

Hypohidrosis (n=135)								
Any infection	270	1856	14545	767	18733	4094	3.54 (2.60–4.82)	3.48 (2.60–4.65)
GI tract infections	7	1825	384	52	18655	279	1.37 (0.63–2.98)	1.44 (0.66–3.13)
Abscesses	11	1825	603	88	18678	471	1.29 (0.61–2.76)	1.15 (0.52–2.52)
Septicaemia	9	1825	493	11	18655	59	7.99 (2.60–24.60)	7.36 (3.06–17.72)
Skin and SC infections	22	1827	1204	106	18665	568	2.06 (1.22–3.48)	2.08 (1.23–3.52)
Viral infections	30	1845	1626	85	18674	455	3.38 (1.94–5.90)	3.29 (1.98–5.47)
Fungal infections	7	1825	384	6	18655	32	13.49 (5.01–36.31)	17.82 (3.84–82.63)
Eye and ear infections	22	1827	1204	106	18665	568	2.06 (1.22–3.48)	2.08 (1.23–3.52)
Pneumonia	54	1836	2941	68	18658	364	7.87 (4.94–12.55)	8.26 (5.11–13.34)
URT infections	24	1841	1303	108	18669	579	2.23 (1.32–3.77)	2.13 (1.25–3.63)
Teeth and oral infections	20	1825	1096	11	18667	59	21.83 (10.92–43.62)	26.40 (10.66–65.35)
Intraabdominal infections	8	1837	435	72	18659	386	1.11 (0.51–2.40)	1.12 (0.54–2.34)
Urinary tract infections	5	1825	274	53	18655	284	0.92 (0.31–2.78)	0.86 (0.28–2.69)
Hyperthermia	11	1825	603	41	18658	220	2.62 (0.88–7.78)	2.69 (1.06–6.83)
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Incontinentia pigmenti (n=75)								
Any infection	127	929	13674	468	9187	5094	2.70 (2.06–3.55)	2.75 (2.09–3.62)
GI tract infections	10	926	1080	38	9143	416	2.68 (1.32–5.47)	2.78 (1.34–5.76)
Abscesses	7	926	756	25	9143	273	2.81 (1.18–6.71)	2.87 (1.15–7.14)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Skin and SC infections	17	926	1836	40	9143	437	4.21 (2.06–8.64)	4.56 (2.13–9.78)
Viral infections	16	926	1728	75	9159	819	2.04 (1.08–3.85)	2.21 (1.18–4.14)
Fungal infections	3	926	324	6	9143	66	5.84 (1.62–21.11)	9.01 (2.21–36.78)
Eye and ear infections	17	926	1836	40	9143	437	4.21 (2.06–8.64)	4.56 (2.13–9.78)
Pneumonia	8	926	864	29	9144	317	2.74 (0.96–7.80)	2.78 (0.92–8.35)
URT infections	18	926	1944	122	9145	1334	1.48 (0.83–2.63)	1.50 (0.88–2.55)
Teeth and oral infections	5	926	540	8	9143	87	5.94 (2.04–17.35)	5.69 (1.68–19.24)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	18	926	1944	36	9143	394	4.84 (2.36–9.96)	4.39 (1.95–9.88)
Hyperthermia	7	926	756	29	9158	317	2.38 (0.92–6.16)	3.22 (1.33–7.83)
Febrile convulsions	8	926	864	13	9143	142	6.15 (2.07–18.33)	6.41 (1.93–21.25)
XLHED, males (n=50)								
Any infection	86	827	10397	324	7904	4099	2.55 (1.75–3.71)	2.49 (1.70–3.64)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	4	827	484	43	7888	545	0.87 (0.19–3.97)	0.79 (0.15–4.06)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	8	827	967	60	7888	761	1.25 (0.57–2.76)	1.23 (0.55–2.77)
Viral infections	10	827	1209	43	7877	546	2.19 (1.05–4.56)	2.18 (1.04–4.54)
Fungal infections	N/A	N/A	N/A	<3	N/A	N/A	N/A	N/A
Eye and ear infections	8	827	967	60	7888	761	1.25 (0.57–2.76)	1.23 (0.55–2.77)
Pneumonia	27	827	3264	24	7877	305	10.80 (5.38–21.67)	10.57 (5.26–21.23)

URT infections	11	827	1330	53	7877	673	2.00 (0.86–4.67)	2.09 (0.90–4.82)
							19.59 (7.74–	
Teeth and oral infections	10	827	1209	5	7877	63	49.55)	35.76 (11.56–110.65)
Intraabdominal infections	3	827	363	26	7877	330	1.09 (0.27–4.36)	1.15 (0.28–4.77)
Urinary tract infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
XLHED, females (n=50)								
Any infection	67	554	12085	250	5626	4444	2.36 (1.57–3.56)	2.28 (1.46–3.55)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Septicaemia	3	551	544	3	5590	54	4.63 (1.45–14.80)	10.07 (2.00–50.62)
Skin and SC infections	3	553	542	34	5590	608	0.80 (0.26–2.44)	0.60 (0.21–1.70)
Viral infections	9	553	1626	21	5596	375	3.96 (1.39–11.26)	4.27 (1.28–14.30)
Fungal infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	3	553	542	34	5590	608	0.80 (0.26–2.44)	0.60 (0.21–1.70)
Pneumonia	11	551	1996	22	5593	393	4.20 (1.93–9.17)	4.77 (1.95–11.66)
URT infections	6	551	1089	26	5605	464	2.28 (0.84–6.18)	2.40 (0.80–7.19)
Teeth and oral infections	3	551	544	5	5602	89	7.12 (1.91–26.52)	7.57 (1.23–46.70)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	11	551	1996	28	5590	501	3.01 (1.35–6.71)	2.99 (1.27–7.08)

Hyperthermia	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hypohidrotic ED, no genetic diagnosis (n=55)								
Any infection	84	762	11024	270	7957	3393	3.31 (2.13–5.13)	3.23 (2.04–5.12)
GI tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Abscesses	3	734	409	37	7941	466	0.91 (0.30–2.75)	0.80 (0.25–2.51)
Septicaemia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	6	734	818	36	7941	453	1.84 (0.67–5.00)	1.75 (0.68–4.49)
Viral infections	14	751	1864	21	7954	264	6.76 (3.09–14.79)	5.24 (2.53–10.86)
Fungal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	6	734	818	36	7941	453	1.84 (0.67–5.00)	1.75 (0.68–4.49)
Pneumonia	17	745	2282	24	7941	302	7.55 (3.36–16.99)	6.82 (2.93–15.89)
URT infections	5	750	666	36	7943	453	1.48 (0.58–3.81)	1.61 (0.64–4.07)
Teeth and oral infections	7	734	954	3	7941	38	34.21 (8.31–140.85)	56.67 (9.40–341.74)
Intraabdominal infections	4	746	536	29	7941	365	1.43 (0.45–4.60)	2.03 (0.86–4.81)
Urinary tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hidrotic ED, no genetic diagnosis (n=55)								
Any infection	30	531	5646	203	5456	3720	1.60 (0.89–2.89)	1.57 (0.89–2.80)
GI tract infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A

Abscesses	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Septicaemia	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Skin and SC infections	7	531	1317	25	5456	458	2.66 (0.77–9.22)	2.67 (0.85–8.36)
Viral infections	5	531	941	22	5456	403	2.41 (0.83–6.98)	2.58 (0.94–7.07)
Fungal infections	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Eye and ear infections	7	531	1317	25	5456	458	2.66 (0.77–9.22)	2.67 (0.85–8.36)
Pneumonia	3	531	565	6	5456	110	4.65 (1.09–19.81)	3.33 (0.71–15.60)
URT infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Teeth and oral infections	3	531	565	4	5456	73	7.39 (2.08–26.29)	8.44 (2.17–32.85)
Intraabdominal infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Urinary tract infections	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Hyperthermia	<3	N/A	N/A	N/A	N/A	N/A	N/A	N/A
Febrile convulsions	0	N/A	N/A	N/A	N/A	N/A	N/A	N/A

^a The number of events for individual infections do not add up to the totals, as people may have multiple infections.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Adjusted for educational level.

Abbreviations: CI, confidence interval; ED, ectodermal dysplasia; GI, gastrointestinal; HR, hazard ratio; IR, incidence rate; SC, subcutaneous; URT, upper respiratory tract; XLHED, X-linked hypohidrotic ectodermal dysplasia.

Table SXIII. Recurrent prescriptions of antimicrobials after ectodermal dysplasia diagnosis

Group	Patients with ED (n=396)			Comparators (n=3960)			HR (95% CI)	
	Events ^a	Person-years at risk	IR per 100,000 person years	Events	Person-years at risk	IR per 100,000 person years	Unadjusted ^b	Adjusted ^c
Any ED (n=396)								
Systemic antibiotics, any	2875	4684	61385	16519	47124	35054	1.75 (1.57–1.95)	1.73 (1.56–1.92)
Phenoxymethylpenicillin	1150	4674	24602	7141	46884	15231	1.62 (1.45–1.81)	1.61 (1.44–1.79)
Aminopenicillins	593	4647	12762	2216	46648	4750	2.68 (2.17–3.32)	2.69 (2.19–3.32)
Macrolides	606	4655	13018	2751	46634	5899	2.21 (1.67–2.94)	2.17 (1.66–2.85)
Cystitis antibiotics	394	4650	8474	3029	46725	6483	1.29 (1.00–1.67)	1.28 (0.99–1.65)
BL resistant penicillins	362	4633	7813	1392	46598	2987	2.62 (2.05–3.35)	2.54 (2.02–3.19)
Quinolones	120	4635	2589	377	46523	810	3.08 (2.03–4.65)	3.03 (2.07–4.45)
Systemic antimycotics	278	4635	5997	1524	46667	3266	1.84 (1.26–2.68)	1.80 (1.25–2.59)
Systemic antivirals	280	4627	6052	1249	46581	2681	2.23 (1.27–3.92)	2.29 (1.33–3.96)
Ophthalmic antimicrobials	868	4662	18617	3912	46686	8379	2.22 (1.82–2.70)	2.21 (1.82–2.68)
Otological antimicrobials	219	4637	4723	718	46520	1543	3.07 (2.26–4.18)	3.10 (2.28–4.20)
Hypohidrosis (n=135)								
Systemic antibiotics, any	1324	1865	70979	6198	18958	32693	2.16 (1.83–2.55)	2.13 (1.81–2.50)
Phenoxymethylpenicillin	480	1857	25844	2872	18881	15211	1.70 (1.44–2.01)	1.68 (1.42–1.98)
Aminopenicillins	350	1835	19069	862	18742	4599	4.11 (3.04–5.55)	4.03 (3.02–5.40)

Macrolides	340	1844	18440	1025	18737	5470	3.41 (2.28–5.10)	3.29 (2.26–4.79)
Cystitis antibiotics	106	1842	5754	849	18721	4535	1.25 (0.64–2.44)	1.23 (0.66–2.29)
BL resistant penicillins	185	1825	10137	549	18713	2934	3.47 (2.51–4.80)	3.47 (2.52–4.79)
Quinolones	51	1833	2782	149	18667	798	3.33 (2.07–5.36)	3.21 (2.04–5.05)
Systemic antimycotics	131	1834	7143	455	18718	2431	3.11 (1.66–5.83)	3.13 (1.84–5.33)
Systemic antivirals	113	1825	6192	507	18723	2708	2.15 (0.92–5.02)	2.55 (1.11–5.83)
Ophthalmic antimicrobials	326	1846	17661	1595	18746	8508	2.06 (1.39–3.05)	2.05 (1.44–2.91)
Otological antimicrobials	72	1835	3923	277	18680	1483	2.63 (1.68–4.12)	2.55 (1.64–3.94)
Incontinentia pigmenti (n=75)								
Systemic antibiotics, any	529	928	57011	3800	9279	40954	1.42 (1.15–1.74)	1.41 (1.16–1.73)
Phenoxymethylpenicillin	214	928	23063	1543	9217	16741	1.38 (1.07–1.79)	1.38 (1.07–1.78)
Aminopenicillins	99	926	10694	667	9210	7242	1.48 (1.07–2.04)	1.47 (1.07–2.02)
Macrolides	80	928	8624	608	9176	6626	1.30 (0.81–2.09)	1.37 (0.87–2.14)
Cystitis antibiotics	134	926	14475	773	9220	8384	1.82 (1.25–2.65)	1.84 (1.27–2.66)
BL resistant penicillins	44	926	4753	278	9170	3032	1.60 (1.03–2.48)	1.54 (0.98–2.41)
Quinolones	12	926	1296	71	9163	775	1.69 (0.76–3.80)	1.63 (0.70–3.81)
Systemic antimycotics	44	926	4753	377	9208	4094	1.18 (0.61–2.27)	1.32 (0.72–2.43)
Systemic antivirals	21	926	2268	214	9150	2339	0.97 (0.36–2.62)	0.96 (0.35–2.63)
Ophthalmic antimicrobials	164	928	17680	992	9191	10793	1.63 (1.31–2.03)	1.62 (1.29–2.03)
Otological antimicrobials	42	926	4537	189	9165	2062	2.18 (1.14–4.20)	2.21 (1.16–4.22)
XLHED, males (n=50)								

Systemic antibiotics, any	583	827	70481	2310	8017	28814	2.46 (1.91–3.17)	2.34 (1.83–2.99)
Phenoxymethylpenicillin	223	827	26959	1239	7987	15513	1.74 (1.38–2.20)	1.70 (1.34–2.16)
Aminopenicillins	168	827	20310	385	7913	4865	4.18 (2.67–6.55)	3.96 (2.60–6.03)
Macrolides	188	827	22728	398	7904	5036	4.46 (2.45–8.10)	4.02 (2.34–6.94)
Cystitis antibiotics	3	827	363	47	7890	596	0.63 (0.15–2.75)	0.60 (0.10–3.47)
BL resistant penicillins	89	827	10760	236	7912	2983	3.61 (2.25–5.78)	3.55 (2.18–5.77)
Quinolones	13	827	1572	47	7877	597	2.49 (1.14–5.44)	2.23 (1.00–4.97)
Systemic antimycotics	4	827	484	45	7905	569	0.84 (0.30–2.38)	0.97 (0.37–2.51)
								4.38 (1.39–
Systemic antivirals	34	827	4110	227	7932	2862	1.32 (0.35–4.95)	13.87)
Ophthalmic antimicrobials	112	827	13540	680	7939	8565	1.60 (1.13–2.25)	1.59 (1.12–2.25)
Otological antimicrobials	36	827	4352	111	7900	1405	3.20 (1.65–6.20)	3.27 (1.70–6.31)
XLHED, females (n=50)								
Systemic antibiotics, any	334	567	58885	2442	5703	42820	1.35 (1.04–1.76)	1.31 (1.00–1.70)
Phenoxymethylpenicillin	147	567	25916	963	5675	16969	1.56 (1.16–2.09)	1.49 (1.12–1.98)
Aminopenicillins	29	551	5262	234	5620	4164	1.28 (0.77–2.15)	1.27 (0.74–2.20)
Macrolides	63	556	11328	390	5624	6934	1.65 (0.84–3.22)	1.59 (0.80–3.16)
Cystitis antibiotics	68	558	12189	678	5636	12029	0.96 (0.55–1.67)	0.91 (0.55–1.53)
BL resistant penicillins	24	551	4354	187	5610	3333	1.32 (0.75–2.32)	1.27 (0.73–2.22)
Quinolones	20	559	3577	68	5590	1217	2.65 (1.32–5.35)	2.72 (1.29–5.75)
Systemic antimycotics	50	553	9039	263	5622	4678	2.07 (0.87–4.92)	1.55 (0.69–3.50)

Systemic antivirals	38	551	6894	147	5594	2628	2.59 (1.12–6.00)	2.60 (1.04–6.48)
Ophthalmic antimicrobials	42	563	7461	445	5614	7926	0.95 (0.64–1.42)	0.93 (0.62–1.38)
Otological antimicrobials	6	551	1089	62	5600	1107	1.03 (0.41–2.56)	1.12 (0.47–2.68)
Hypohidrotic ED, no genetic diagnosis (n=55)								
Systemic antibiotics, any	596	758	78609	2702	8034	33631	2.28 (1.73–3.02)	2.19 (1.71–2.80)
Phenoxymethylpenicillin	232	750	30928	1179	8005	14729	2.11 (1.59–2.79)	2.01 (1.56–2.58)
Aminopenicillins	129	744	17332	294	7950	3698	4.63 (2.72–7.88)	4.42 (2.62–7.43)
Macrolides	98	748	13107	513	7971	6436	2.05 (1.05–3.99)	1.95 (1.05–3.63)
Cystitis antibiotics	87	744	11687	487	7974	6107	1.74 (0.82–3.67)	1.57 (0.83–2.97)
BL resistant penicillins	90	734	12263	216	7950	2717	4.51 (2.56–7.94)	3.72 (2.49–5.57)
Quinolones	45	734	6132	65	7954	817	7.23 (3.27–15.98)	5.89 (3.32–10.44)
Systemic antimycotics	75	741	10124	276	7960	3468	2.77 (1.49–5.14)	2.63 (1.48–4.67)
Systemic antivirals	22	734	2998	267	7948	3359	0.84 (0.27–2.61)	0.94 (0.31–2.83)
Ophthalmic antimicrobials	96	743	12919	551	7955	6926	1.86 (1.19–2.89)	1.83 (1.22–2.75)
Otological antimicrobials	27	744	3629	92	7941	1159	3.09 (1.42–6.71)	3.08 (1.56–6.07)
Hidrotic ED, no genetic diagnosis (n=55)								
Systemic antibiotics, any	220	538	40877	1796	5500	32655	1.26 (0.97–1.65)	1.27 (0.97–1.66)
Phenoxymethylpenicillin	96	538	17850	755	5490	13752	1.33 (0.98–1.81)	1.34 (0.99–1.82)
Aminopenicillins	33	537	6143	156	5457	2858	2.23 (1.27–3.94)	2.25 (1.30–3.91)
Macrolides	51	531	9599	331	5458	6064	1.63 (0.71–3.74)	1.61 (0.72–3.58)

Cystitis antibiotics	27	537	5027	410	5473	7492	0.68 (0.37–1.25)	0.74 (0.42–1.32)
BL resistant penicillins	19	531	3576	149	5464	2727	1.28 (0.61–2.68)	1.27 (0.62–2.60)
Quinolones	5	531	941	39	5456	715	1.23 (0.48–3.14)	1.10 (0.47–2.61)
Systemic antimycotics	7	531	1317	183	5474	3343	0.39 (0.11–1.38)	0.35 (0.09–1.36)
Systemic antivirals	18	531	3388	126	5460	2308	1.88 (0.66–5.30)	1.74 (0.67–4.53)
Ophthalmic antimicrobials	60	535	11207	373	5467	6822	1.62 (1.12–2.33)	1.69 (1.16–2.45)
Otological antimicrobials	10	531	1882	57	5456	1045	1.81 (0.72–4.53)	2.69 (1.16–6.21)

^a The number of events for individual prescriptions do not add up to the totals, as people may have multiple categories of prescriptions.

^b Adjusted for matching factors (age, sex, and municipality) by design.

^c Adjusted for educational level.

Abbreviations: BL, beta-lactamase; CI, confidence interval; ED, ectodermal dysplasia; HR, hazard ratio; IR, incidence rate; XLHED, X-linked hypohidrotic ectodermal dysplasia.