

Table 1. Description of the 74 deaths caused by malignant neoplasms in Down syndrome population, California, January 1, 1988 – December 31, 2002

No.	Sex	Age at death (years)	Year of death	Malignant neoplasm (ICD-9)
1	Female	47.1	1989	Long bones of lower limb (170.7)
2	Female	3.7	1989	Unspecified leukemia (208.9)
3	Female	56.2	1989	Acute leukemia NOS (208.0)
4	Female	2.3	1990	Megakaryocytic leukemia (207.2)
5	Female	2.7	1990	Unspecified leukemia (208.9)
6	Female	3.0	1990	Acute leukemia NOS (208.0)
7	Male	36.0	1990	Scalp and skin of neck (173.4)
8	Male	9.4	1991	Hodgkin's disease, unspecified (201.9)
9	Female	12.3	1991	Acute lymphoid leukemia (204.0)
10	Male	63.2	1991	Bladder, part unspecified (188.9)
11	Female	2.2	1992	Acute leukemia NOS (208.0)
12	Male	10.4	1992	Acute lymphoid leukemia (204.0)
13	Female	12.1	1992	Acute lymphoid leukemia (204.0)
14	Male	20.7	1992	Acute lymphoid leukemia (204.0)
15	Male	23.2	1992	Soft tissue, NOS (171.9)
16	Female	28.1	1992	Acute lymphoid leukemia (204.0)
17	Male	30.0	1992	Lymphoma NOS (202.8)

18	Female	48.4	1992	Breast, unspecified (174.9)
19	Male	3.4	1993	Acute lymphoid leukemia (204.0)
20	Female	11.0	1993	Acute leukemia NOS (208.0)
21	Male	3.4	1994	Acute lymphoid leukemia (204.0)
22	Female	4.1	1994	Acute leukemia NOS (208.0)
23	Male	14.7	1994	Acute lymphoid leukemia (204.0)
24	Male	17.3	1995	Unspecified leukemia (208.9)
25	Male	26.1	1995	Acute leukemia NOS (208.0)
26	Male	49.5	1995	Testis NOS (186.9)
27	Female	10.9	1996	Acute lymphoid leukemia (204.0)
28	Male	37.6	1996	Liver, primary (155.0)
29	Male	51.6	1997	Lymphoma NOS (202.8)
30	Female	3.9	1998	Acute leukemia NOS (208.0)
31	Female	7.3	1998	Unspecified leukemia (208.9)
32	Male	7.8	1998	Acute lymphoid leukemia (204.0)
33	Male	9.7	1998	Acute lymphoid leukemia (204.0)
34	Male	12.3	1998	Acute lymphoid leukemia (204.0)
35	Male	20.8	1998	Other, without specification of site (199.1)
36	Male	49.6	1998	Cerebrum (191.0)
37	Male	7.7	1999	Acute lymphoid leukemia (204.0)
38	Female	46.1	1999	Breast, unspecified (174.9)
39	Male	50.9	1999	Brain, unspecified (191.9)

40	Male	58.8	1999	Lymphoma NOS (202.8)
41	Female	7.3	2000	Acute lymphoid leukemia (204.0)
42	Male	12.5	2000	Soft tissue, NOS (171.9)
43	Female	19.1	2000	Other, without specification of site (199.1)
44	Male	21.4	2000	Lymphoma NOS (202.8)
45	Female	22.1	2000	Acute lymphoid leukemia (204.0)
46	Male	22.7	2000	Brain, unspecified (191.9)
47	Male	26.0	2000	Bronchus and Lung NOS (162.9)
48	Male	43.0	2000	Liver cell (155.0)
49	Female	53.4	2000	Liver (155)
50	Female	54.6	2000	Meninges (192.1)
51	Female	3.3	2001	Acute myeloid leukemia
52	Male	5.7	2001	Acute leukemia NOS (208.0)
53	Female	8.3	2001	Lymphosarcoma (200.1)
54	Male	8.8	2001	Acute lymphoid leukemia (204.0)
55	Female	17.4	2001	Acute lymphoid leukemia (204.0)
56	Male	29.6	2001	Acute leukemia NOS (208.0)
57	Male	38.6	2001	Pancreas NOS (157.9)
58	Male	38.8	2001	Liver cell (155.0)
59	Male	41.5	2001	Testis NOS (186.9)
60	Male	44.0	2001	Esophagus NOS (150.9)
61	Male	44.6	2001	Liver cell (155.0)

62	Female	68.1	2001	Bronchus and Lung NOS (162.9)
63	Male	76.5	2001	Bronchus and Lung NOS (162.9)
64	Female	4.5	2002	Acute myeloid leukemia
65	Female	7.9	2002	Acute lymphoid leukemia (204.0)
66	Male	10.3	2002	Acute lymphoid leukemia (204.0)
67	Female	13.3	2002	Acute lymphoid leukemia (204.0)
68	Female	16.6	2002	Bone and cartilage NOS (170.9)
69	Female	34.6	2002	Acute lymphoid leukemia (204.0)
70	Male	35.8	2002	Acute lymphoid leukemia (204.0)
71	Male	38.2	2002	Liver NOS (155.2)
72	Female	41.4	2002	Other, without specification of site (199.1)
73	Female	44.6	2002	Acute lymphoid leukemia (204.0)
74	Female	52.0	2002	Skin NOS (173.9)

Table 2. Standardized mortality ratios for cancer among persons with Down syndrome, California, January 1, 1988 – December 31, 2002.

Cause of death (ICD-9)	Deaths			95% C.I.§
	Obs*	Exp†	SMR‡	
All malignant neoplasms	74	28.84	2.6	2.0-3.2
Lip, oral cavity, pharynx (140-149)	0	0.58	0.0	0.0-6.4
Digestive organs and peritoneum (150-159)	8	5.70	1.4	0.6-2.8
Colon (153)	1	1.64	0.6	0.0-3.4
Liver (155)	5	0.89	5.6	1.8-13.1
Gallbladder (156)	0	0.14	0.0	0.0-26.4
Respiratory and intrathoracic organs (160-165)	3	6.07	0.5	0.1-1.4
Nasal, larynx (160-161)	0	0.20	0.0	0.0-18.6
Trachea, bronchus, lung (162)	3	6.00	0.5	0.1-1.5
Bone, connective tissue, skin, breast (170-175)	8	5.20	1.5	0.7-3.0
Bone and cartilage (170)	2	0.33	6.0	0.7-21.7
Skin (172-173)	2	0.89	2.3	0.3-8.1

Breast (174-175)	2	3.42	0.6	0.1-2.1
Genitourinary organs (179-189)	3	3.15	1.0	0.2-2.8
Testis (186)	2	0.16	12.5	1.5-45.1
Ovary and other uterine adnexa (183)	0	0.79	0.0	0.0-4.7
Bladder (188)	1	0.25	3.9	0.1-21.9
Malignant neoplasms of other and unspecified sites (190-199)	8	3.90	2.1	0.9-4.0
Brain (191)	4	1.8	2.2	0.6-5.7
Without specification of site (199)	3	1.45	2.1	0.4-6.0
Malignant neoplasms of lymphatic and hematopoietic tissue (200-208)	44	4.26	10.3	7.5-13.9
Lymphomas (200-202)	6	1.64	3.7	1.3-8.0
Leukemias (204-208)	38	2.26	16.8	11.9-23.1
Lymphoid leukemia (204)	23	0.83	27.6	17.5-41.4
Myeloid leukemia (205)	2	0.92	2.2	0.3-7.9
Other specified leukemia (207)	1	0.02	51.1	1.3-285.0
Leukemia of unspecified cell type (208)	12	0.47	25.4	13.1-

Tobacco related cancers 	13	12.24	1.1	0.6-1.8
All malignant neoplasms other than those of lymphatic and hematopoietic tissue (i.e., excluding 200-208)	30	24.58	1.2	0.8-1.7

* Obs = number of observed deaths among 14,781 persons with Down syndrome in California, 1988-2002.

† Exp = expected numbers of deaths based on age-adjusted cause-specific mortality rates in the California general population, 1988-2002.

‡ Ratio of observed to expected number of deaths, obs/exp.

§ Confidence interval based on the assumption that the observed deaths follow a Poisson distribution.(20)

|| Tobacco related cancers from Monograph 83 of the IARC on tobacco smoking(38): lung, trachea, bronchus (ICD9 codes 162), oral cavity (140-145), pharynx (oro-, naso-, hypo-) (146-148), larynx (161), oesophagus (150), pancreas (157), bladder and renal pelvis (188), nasal cavities and sinuses (160), stomach (151), liver (155), kidney (189), uterine cervix (180), myeloid leukemia (205).

Table 3. Comparison of SMRs and SIRs between the present study and selected other studies

Cause of death (ICD-9 codes)	Present study		Hill et al. 2003 [3]	Hermon et al. 2001 [6]	Oster et al. 1975 [7]	Holland et al. 1962 [25]	Boker et al. 2001, 2002 [4,5]	Hasle et al. 2000 [10]	Goldacre et al. 2004	Patja et al. 2006	Sullivan et al. 2007		
	Obs	SMR (95% CI)	SMR (95% CI)	SIR (95% CI)	SMR (95% CI)	SIR (95% CI)	SMR (95% CI)	SMR (95% CI)	SIR (95% CI)	SIR (95% CI)	SIR (95% CI)		
All malignant neoplasms	74	2.6 (2.0-3.2)	3.9 (1.9-5.0)	1.7 (1.3-2.1)	1.87 (1.21-2.76)	1.27 (0.86-1.82)	3.39§ (NA) 1.35 (NA)	---	1.33¶ (0.77-2.12)	1.20 (0.92-1.55)	2.7 (1.80-3.90)	0.9 (0.7-1.1)	1.1 (0.68-1.68)
Lip, oral cavity, pharynx (140-149)	0	0.0 (0.0-6.4)	---	---	---	---	---	---	---	0.00 (0.00-2.88)	---	0.0 (0.0-2.6)	---
Digestive organs and peritoneum (150-159)	8	1.4 (0.6-2.8)	---	---	---	---	---	---	---	0.61 (0.17-1.57)	---	1.3 (0.6-2.2)	---
Colon (153)	1	0.6 (0.0-3.4)	3.3 (0.7-9.6)	2.1 (0.6-5.3)	---	---	---	---	---	0.89 (0.10-3.23)	3.1 (0.4-11.1)	1.5 (0.4-3.9)	---
Liver (155)	5	5.6 (1.8-13.1)	7.2 (0.9-26.1)	6.0 (1.2-17.5)	---	---	---	---	---	---	---	2.4 (0.1-13.2)	---
Gallbladder (156)	0	0.0 (0.0-26.4)	8.2 (1.0-29.6)	---	---	---	---	---	---	---	---	6. (0.7-21.6)	13.56 (0.34-75.6)
Respiratory and intrathoracic organs (160-165)	3	0.5 (0.0-1.4)	---	---	---	---	---	---	---	0.20 (0.00-1.12)	---	0.0 (0.0-0.9)	---
Nasal, larynx (160-161)	0	0.0 (0.0-18.6)	---	---	---	---	---	---	---	---	---	---	---

Trachea, bronchus, lung (162)	3	0.5 (0.0.1-1.5)	---	---	---	---	---	---	---	0.24 (0.00-1.32)			
Bone, connective tissue, skin, breast (170-176)	8	1.5 (0.7-3.0)	---	---	---	---	---	---	---	---			
Bone and cartilage (170)	2	6.0 (0.7-21.7)	---	---	---	---	---	---	---	---		2.1 (0.1-11.9)	
Skin (172-173)	2	0.89 (0.3-8.1)	---	---	---	---	---	---	---	0.25 (0.03-0.89)			
Breast (174-175)	2	0.6 (0.1-2.1)	---	0.5 (0.1-1.4)	0.62 (NS)	---	---	---	---	0.00 (0.00-0.41)		0.4 (0.1-0.8)	
Genitourinary organs (179-189)	3	1.0 (0.2-2.8)	---	---	---	---	---	---	---	---			
Testis (186)	2	12.5 (1.5-45.1)	25.2 (3.0-90.9)	3.7 (1.0-9.4)	8.40 (NS)	3.46 (NA)	---	---	---	1.86 (0.50-4.77)	12.0 (2.5-35.6)	4.8 (1.8-10.4)	1.94 (0.05-10.83)
Ovary and other uterine adnexa (183)	0	0.0 (0.0-4.7)			4.05 (NS)	3.83 (NA)	---	---	---	1.97 (0.40-5.77)		0.5 (0.0-3.0)	
Bladder (188)	1	3.9 (0.1-21.9)	---	---	---	---	---	---	---	1.69 (0.34-4.93)		0.0 (0.0-2.9)	

	Present study	Hill et al. 2003	Hermon et al. 2001	Oster et al. 1975	Holland et al. 1962	Boker et al. 2001, 2002	Hasle et al. 2000	Goldacre et al. 2004	Patja et al. 2006	Sullivan et al. 2007		
Cause of death (ICD-9 codes) - Continued	Obs	SMR (95% CI)	SMR (95% CI)	SIR (95% CI)	SMR (95% CI)	SIR (95% CI)	SMR (95% CI)	SMR (95% CI)	SIR (95% CI)	SIR (95% CI)	SIR (95% CI)	SIR (95% CI)
Malignant neoplasms of other and unspecified sites (190-199)	8	2.1 (0.9-4.0)	---	---	---	---	---	---	---	---	---	---
Brain (191)	4	2.2 (0.6-5.7)	---	0.7 (0.1-2.4)	---	---	---	---	---	0.30 (0.00-1.68)	0.4 (0.0-1.3)	1.6 (0.04-8.92)
Without specification of site (199)	3	2.1 (0.4-6.0)	---	0.6 (0.1-3.2)	---	---	---	---	---	3.27 (0.66-9.56)	0.0 (0.0-4.5)	---
Malignant neoplasms of lymphatic and hematopoietic tissue (200-208)	44	10.3 (7.5-13.9)	---	9.1 (6.4-12.4)	---	---	---	---	---	---	---	---
Lymphomas (200-202)	6	3.7 (1.3-8.0)	3.9* (0.5-14.2)	1.0* (0.1-3.5)	2.08* (NS)	---	---	---	---	0.00* (0.00-2.13)	2.7 (0.3-9.6)	0.6 (0.1-2.3)
Lymphoid leukemia (204)	23	27.6 (17.5-41.4)	33.2† (23.1- 46.2)	24.2 (15.2- 36.6)	13.04† (6.51- 23.34)	17.26† (0.96- 29.61)	2† cases significantly more than expected	17.95† (S)	6.7¶ (0.7-24.2)	24.36 (14.9-37.6)	22.2 (10.9-40.6)	60.0** (12.1-175.3)
Myeloid leukemia (205)	2	2.2 (0.3-7.9)	---	28.2† (15.7- 48.3)	---	---	---	---	3.0¶ (0.0-16.6)	20.28 (10.5-35.4)	17.2 (5.5-40.9)	4.90** (0.06-27.27)
Other specified leukemia (207)	1	51.1 (1.3-285.0)	---	---	---	---	---	---	---	---	---	---

Leukemia of unspecified cell type (208)	12	25.4 (13.1-44.3)	---	33.3 (0.8-185.6)	---	---	---	---	12.0 ¶(0.2-67.0)	1.93 (0.03-10.8)
									130.4 ** (26.2-381.1)	
Tobacco related cancers	13	1.1 (0.6-1.8)	---	---	---	---	---	---	---	---

Obs. Number of observed deaths among the Down syndrome population, California, January 1, 1988 – December 31, 2002

* Non Hodgkin lymphomas only (202)

† All leukemia (204-208)

‡ Nonlymphocytic acute leukemia

§ Period 1949-1959

|| Period 1960-1971

¶ Period before 1979

** Period 1979-1995

NS. Not significant ($p > 0.05$), exact p-value not given

S. Significant ($p < 0.05$), exact p-value not given

NA. Not available

