National Registry of Childhood Tumours

Progress Report, 2012

The contents of this report follow a scheme that was approved by the NCIN Children, Teenagers and Young Adults Clinical Reference Group (CTYA CRG) in 2010.

1. Registration and Follow-up

The NRCT is population-based for cases of cancer diagnosed among children aged under 15 years in Great Britain (England, Scotland and Wales) from 1962 onwards. Since 1993 ascertainment of cases in Northern Ireland has also been virtually complete, hence the NRCT is population-based for the whole of the UK from 1993 onwards. Cases are ascertained from national and regional cancer registries throughout the UK, from specialist children's tumour registries in certain regions of England, from death certificates throughout Great Britain, from entries to clinical trials, and from the paediatric oncology principal treatment centres affiliated to the CCLG throughout the UK. Registration data are also collected from the CCLG centre in Dublin, thus providing complete coverage of CCLG patients throughout the British Isles.

Table 1.1 shows numbers of registered cases of cancer in the NRCT for children resident in Great Britain at diagnosis from 1962 to 2010, the latest year for which registration is virtually complete from all sources. Table 1.2 shows the numbers for the whole of the UK for 1993-2010. The diagnostic categories in Tables 1.1 and 1.2 are the 12 main groups of the International Classification of Childhood Cancer, Third Edition (ICCC-3). The NRCT now contains 68,572 registrations for children with cancer in Great Britain over the 49-year period 1962-2010, and 28,328 registrations for the UK over the 18-year period 1993-2010.

At the time of writing, registration data for over 46,000 CCLG patients aged under 15 at diagnosis and diagnosed during 1977 onwards have been entered into the NRCT database. Throughout this report, numbers of CCLG registrations include registrations for non-malignant neoplasms and allied conditions in addition to those for cancers contained in ICCC-3, except where otherwise stated. Registration of CCLG patients up to 2010 is virtually complete and about 40% of registrations for 2011 have been processed. Table 1.3 shows all registered children under 15 years of age at diagnosis on the database, classified by CCLG centre and year of diagnosis. The centres at Nottingham and Leicester recently amalgamated to form a single East Midlands centre, and there is close co-operation between GOS and UCLH. Throughout this report, data are shown separately for each of these centres. Table 1.4 shows the same patients

classified by detailed diagnostic category and year of diagnosis. Table 1.5(i) shows the numbers of registrations for 2001-2005 by centre and broad diagnostic group. Overall, the two most frequent diagnostic groups were leukaemia (30%) and CNS tumours (22%). A similar pattern was found at most individual centres, but there were several exceptions. At Bart's/Royal London, the equal largest groups were leukaemia and retinoblastoma (36%). CNS tumours outnumbered leukaemia by a considerable margin at Nottingham, but were hardly ever registered from Leicester, Bart's/Royal London and Middlesex/UCLH. Bone tumours were the largest group at Middlesex/UCLH, accounting for 31% of registrations. Table 1.5(ii) shows similar data for 2006-2011. Registration from Bart's/Royal London ceased in 2005. GOS, like Nottingham, had more registrations for CNS tumours than for leukaemia. Otherwise, there was little change from the patterns observed in 2001-2005.

Follow-up information is obtained from matching with population-based death notifications for children dying of neoplasms in Great Britain, from flagging at the NHS Information Centre and (especially for recently diagnosed patients and for those registered in Ireland) from direct enquiry to CCLG centres.

Table 1.1 Numbers of registrations by ICCC-3 main diagnostic group. National Registry of Childhood Tumours, Great Britain, 1962-2010

	1962-2010	1962-1990	1991-2000	2001-2010
Leukaemias, myeloproliferative & myelodysplastic diseases	22143	12669	4695	4779
Lymphomas & reticuloendothelial neoplasms	7043	4032	1425	1586
CNS & miscellaneous intracranial & intraspinal neoplasms	16592	8977	3623	3992
Neuroblastoma & other peripheral nervous cell tumours	4433	2617	896	920
Retinoblastoma	1969	1118	429	422
Renal tumours	3947	2298	798	851
Hepatic tumours	609	295	138	176
Malignant bone tumours	2959	1748	5674	647
Soft tissue & other extarosseous sarcomas	4357	2306	1027	1024
Germ cell tumours, trophoblastic tumours & neoplasms of gonads	2109	1088	491	530
Other malignant epithelial neoplasms & malignant melanomas	2024	1023	479	522
Other & unspecified malignant neoplasms	387	202	93	92
Total	68572	38373	14658	15541

Table 1.2 Numbers of registrations by ICCC-3 main diagnostic group. National Registry of Childhood Tumours, United Kingdom, 1993-2010

	1993-2010	1993-2000	2001-2010
Leukaemias, myeloproliferative & myelodysplastic diseases	8859	3945	4914
Lymphomas & reticuloendothelial neoplasms	2828	1187	1641
CNS & miscellaneous intracranial & intraspinal neoplasms	7208	3084	4124
Neuroblastoma & other peripheral nervous cell tumours	1684	741	943
Retinoblastoma	781	345	436
Renal tumours	1542	664	878
Hepatic tumours	302	116	186
Malignant bone tumours	1137	475	662
Soft tissue & other extarosseous sarcomas	1908	853	1055
Germ cell tumours, trophoblastic tumours & neoplasms of gonads	944	401	543
Other malignant epithelial neoplasms & malignant melanomas	955	417	538
Other & unspecified malignant neoplasms	180	79	101
Total	28328	12307	16021

TABLE 1.3 CCLG REGISTRATIONS FOR CHILDREN AGED UNDER 15, BY CENTRE, 1977-2011

Diag Year

Centre	1977-1978	1979-1980	1981-1982	1983-1984	1985-1986	1987-1988	1989-1990	1991-1992	1993-1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
Aberdeen	1	3	17	25	21	22	36	24	28	15	8	18	21	14	9	11	16	17	15	12	8	10	14	10	10	3	388
Barts/RLH	56	65	73	78	126	130	146	166	161	76	79	79	59	74	67	69	55	51	36	6							1652
Belfast		41	45	55	53	65	78	70	69	29	37	37	35	30	33	29	33	49	31	36	35	36	31	40	48	17	1062
Birmingham	95	160	182	188	221	253	279	301	293	170	151	152	161	144	142	141	170	165	191	158	150	174	168	174	172	96	4651
Bristol	81	101	124	111	123	151	175	146	194	123	114	105	99	110	79	86	103	92	89	89	101	96	113	111	92	34	2842
Cambridge		33	29	39	58	78	96	111	89	60	51	66	73	83	65	71	85	83	99	82	113	101	97	124	125	68	1979
Cardiff	45	57	67	72	59	76	82	81	82	53	45	44	56	71	48	57	56	47	33	62	66	55	73	52	51	16	1506
Dublin		12	81	100	132	139	132	126	163	82	69	79	98	91	100	119	115	120	129	99	113	122	128	142	84		2575
Edinburgh	20	25	24	44	63	64	49	54	70	42	36	53	42	44	47	51	68	49	58	47	44	39	54	48	29	18	1182
Glasgow	69	76	75	118	111	111	130	125	140	63	53	64	63	60	46	62	61	61	68	63	56	64	88	74	74	40	2015
GOS	203	235	309	306	275	282	257	296	381	193	188	210	155	187	157	145	214	176	184	180	197	203	219	183	154	58	5547
Leeds	84	109	109	108	129	150	169	183	187	98	90	91	107	90	97	108	108	92	88	81	97	88	87	89	85	36	2760
Leicester			18	39	51	31	49	36	41	23	19	15	21	15	28	23	26	20	30	23	33	26	24	21	22	5	639
Liverpool	70	94	76	88	101	99	116	124	141	76	89	70	77	85	77	84	100	84	109	90	80	83	83	79	83	49	2307
Manchester	131	183	177	185	158	183	173	182	229	124	120	122	97	122	111	134	117	136	104	123	112	104	120	107	131	48	3533
Middlesex/UCLH						1	4	23	50	25	24	32	46	36	40	30	36	40	44	43	28	39	42	43	36	13	675
Newcastle	55	104	110	110	119	124	120	134	128	69	78	78	71	73	80	91	90	70	85	87	92	65	82	72	73	37	2297
Nottingham	28	23	60	47	65	77	90	94	91	61	47	49	72	65	76	62	52	59	60	46	73	50	67	75	73	41	1603
Oxford				1	2	1	4	32	90	45	52	49	56	57	57	56	55	69	70	77	67	74	71	56	74	25	1140
Royal Marsden	71	99	83	90	80	113	113	129	115	70	83	83	111	90	116	109	125	84	88	128	109	111	151	148	140	60	2699
Sheffield	65	76	77	61	80	80	86	83	86	45	50	50	51	58	59	63	69	58	48	56	63	66	49	57	58	29	1623
Southampton	13	52	59	56	61	77	92	125	129	53	74	70	70	62	59	68	68	73	67	81	61	69	74	70	86	31	1800
Total	1087	1548	1795	1921	2088	2307	2476	2645	2957	1595	1557	1616	1641	1661	1593	1669	1822	1695	1726	1669	1698	1675	1835	1775	1700	724	46475

TABLE 1.4 CCLG REGISTRATIONS FOR CHILDREN AGED UNDER 15, BY DIAGNOSTIC GROUP, 1977-2011

Diag Year

DiagGpText	1977-1978	1979-1980	1981-1982	1983-1984	1985-1986	1987-1988	1989-1990	1991-1992	1993-1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
Precursor-cell ALL	332	485	531	570	577	631	695	711	763	378	379	377	427	389	377	369	454	394	427	399	403	369	414	433	417	152	11853
Mature B-cell leukaemia		7	6	11	14	6	5	16	11	5	5	5	5	5	5	8	7	3	2	1	2	2	2	1	1		135
AML	77	86	108	101	127	125	135	120	155	68	81	75	66	85	69	74	81	89	82	70	76	71	77	87	75	35	2295
CML	4	7	5	10	15	8	15	5	8	5	6	13	4	6	7	4	8	13	6	3	11	13	5	7	4	3	195
MDS	1		3	1	3	6	9	21	11	10	12	7	15	11	5	9	11	3	4	8	3	8	5	5	1		172
JMML/CMML	2	4	8	6	10	7	8	10	7	2	6	10	4	12	6	7	5	9	10	15	4	5	6	4	2	1	170
Other and unspecified leukaemia	5	5	6	3	6	5	8	4	5	6	2	7	5	3		3	5	2	4	4	4	2	5	3	5	3	110
Hodgkin lymphoma	58	78	90	85	110	72	74	105	95	65	57	61	59	64	69	82	100	55	84	82	79	70	80	78	60	43	1955
NHL	84	115	111	117	131	152	154	162	153	93	74	92	103	93	91	79	100	73	83	82	89	82	100	83	81	28	2605
Other lymphoreticular	4	10	5	3	2	2	3	2		1				1			2		1	1	1		1		1	1	41
Ependymoma	15	19	20	27	23	33	38	35	49	30	22	29	32	27	32	27	31	30	32	22	27	37	36	31	29	15	748
Choroid plexus tumours	2		3	7	2	5	8	10	20	7	10	12	6	10	10	11	8	10	12	13	12	10	12	11	10	5	226
Low-grade astrocytoma	33	58	32	60	86	86	115	133	206	128	109	129	128	132	129	166	126	129	128	143	139	146	143	136	121	38	2979
High-grade astrocytoma	6	11	8	20	17	27	38	48	48	34	28	25	26	25	19	22	28	30	23	33	18	24	27	17	27	11	640
Unspecified astrocytoma	11	2	13	8	9	9	7	18	27	6	2	3	4		3	5	6	2	4	3	2	3	4	20	13	7	191
Medulloblastoma	42	58	65	71	59	66	73	98	100	48	52	51	54	56	49	51	64	60	62	54	62	49	47	52	64	26	1533
Other embryonal CNS		1	4	7	14	25	33	35	30	18	19	18	22	25	18	22	19	15	15	23	30	13	21	30	20	10	487
Other glioma	23	25	24	37	38	40	56	53	46	36	30	35	39	37	41	45	35	39	40	27	50	46	41	44	38	16	981
Pituitary adenoma and carcinoma		1	1				2	2	1	4		1		2	2	2	2	4	3	4	6	3	5	2	6		53
Craniopharyngioma	10	6	5	4	9	10	13	22	28	20	21	14	18	18	20	13	19	17	21	19	20	17	27	17	18	8	414
Pineal parenchymal tumours	6	3	6	5	4	6	3	3	10	2	2	5	6	5	4	4	5	4	7	6	6	5	2	3	5	3	120
Neuronal and neuronal-glial tumours	1	1	2	1	1	3	3	8	21	12	14	23	16	15	20	11	19	9	20	32	28	16	27	15	19	13	350
Meningioma	1	2	2		1	2	4	7	9	3	3	4	8		1	2	1	4	3	5	6	2	7	4	2		83
Unspecified CNS tumours	8	4	8	3	7	3	5	5	7	13	2	5	7	6	3	4	8	8	10	8	6	10	13	11	8		172
Neuroblastoma	71	106	162	151	149	210	212	195	176	92	101	97	103	110	78	106	106	121	88	104	81	100	95	101	104	50	3069
Other malignant peripheral nerve cell		2	2	2		3	1	2		2		1	2		3	1	1	1	3		2	2			2		32
Retinoblastoma	5	13	14	42	66	74	75	92	109	39	36	43	31	35	42	35	52	42	43	33	41	46	38	42	25	16	1129
Wilms Tumour	77	97	111	132	130	157	164	146	166	66	83	88	82	66	81	91	98	86	87	83	77	81	83	77	77	46	2532
Rhabdoid renal tumour		2	6	4	3	4	3	5	4	1	4	5	2	3	1	3	4	5	5	3	5	4	1	2	3	1	83
Renal sarcomas	1	3	3	6	6	9	3	4	8	2	2	4	2	3	5	4	5	4	2	3	4	2	5	6	1	2	99
Renal pPNET							1	1	2	2		2			1	2	3	1	1			1			1		18
Renal carcinoma	1	3	1	3		3	1	3	5	1	4			1	2	1	1		4	3	2	1	5	4		1	50
Unspecified malignant renal							1																				1
Hepatoblastoma	11	8	15	13	15	19	14	21	27	8	16	13	9	9	15	20	16	19	16	15	22	19	11	14	15	16	396
Hepatic carcinoma	2	2	3	6	2	7	4	3	4	2	2	2	2	5	2	1	2	2	7	2		4	3		2	2	73
Unspecified malignant hepatic																	1										1
Osteosarcoma	12	36	42	33	38	42	35	39	66	47	36	30	28	34	32	34	44	35	28	34	35	29	54	33	36	18	930
Chondrosarcoma	1	1	1	1	2		1	1		1	1		1	1	2	1	1	1									17
ESFT of bone	17	49	43	50	43	45	48	45	40	27	24	20	21	27	25	37	31	42	27	19	30	22	28	20	24	10	814
Other malignant bone	4	3	3	4	4	1	2	1	3	3		3	2	3	1	3	1	3	1	3		1	2	1	1	3	56
Rhabdomyosarcoma	46	82	100	111	117	124	117	128	134	62	65	55	51	58	55	45	52	71	65	60	38	54	54	62	59	30	1895
Fibrosarcoma, etc.	4	2	15	4	9	7	8	8	4	3	1	5	1	4	4	4	2	1	6	3	4	1	9	6	8	2	125

TABLE 1.4 CCLG REGISTRATIONS FOR CHILDREN AGED UNDER 15, BY DIAGNOSTIC GROUP, 1977-2011

Diag Year

DiagGpText	1977-1978	1979-1980	1981-1982	1983-1984	1985-1986	1987-1988	1989-1990	1991-1992	1993-1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
MPNST	1	1	2	3	1	3	5	5	3	6	3	4	3	6	2	3	1	5	4	2	2	4	3	2	2	1	77
Kaposi sarcoma	1											1										3					5
Extraosseous ESFT	2	7	4	8	10	15	21	29	30	16	14	23	17	13	15	21	19	18	19	10	15	11	15	17	16	5	390
Extrarenal rhabdoid tumour					2	2	3	3	6	3	3	2		3		2		2	1	3	3	6	8	11	4	5	72
Fibrohistiocytic sarcomas	1	1	5		1	3	3	2	4	1			2	2	1	2	3		1	2			3	4	4		45
Synovial sarcoma		3	2	2	3	6	5	6	12	9	1	5	6	9	5	5	7	4	8	4	8	7	7	6	5	2	137
Other specified soft-tissue sarcomas	5	9	4	6	7	9	5	10	13	11	3	7	1	4	8	11	7	4	8	5	11	5	8	2	1	3	167
Unspecified soft-tissue sarcoma	8	8	2	11	6	11	10	6	5	9	6	9	3	4	4	8	6	6	7	5	3	7	7	5	5	2	163
Intracranial & intraspinal germ cell tumours	6	10	9	9	16	9	12	35	29	9	20	9	11	18	16	16	15	19	17	14	16	22	13	18	17	7	392
Other malignant extragonadal germ cell tumours	11	8	18	10	11	26	24	14	22	9	13	7	11	12	12	14	15	11	15	17	17	13	16	15	16	5	362
Gonadal germ cell tumours	18	23	27	30	46	37	26	26	35	26	21	22	14	21	22	20	14	12	14	17	19	23	24	19	23	5	584
Other malignant gonadal tumours			2	1	3	5	2		1		1			1	1	1		1			2	2	2	1			26
Adrenocortical carcinoma	4	3	4	3	3	1	1	6	2	2	2	2	4	2	5	7	1	3	2	1	3	1		1			63
Thyroid carcinoma	1	4	1	3	4	4	4	3	3	2	7	2	7	5	7	13	6	6	9	11	8	11	10	10	14		155
Nasopharyngeal carcinoma	4	6	7	5	5	5	1	7	3	3	4	2	3	3	3	1		5	4	1	5	4	2	3	1		87
Malignant melanoma	2	6	2	4	3	4	1	7	10	7	4	3	3	7	4	3	9	3	6	3	4	7	5	3	1		111
Skin carcinoma	1	1	1		1				2			1	2		1			1		2	1	1			2	1	18
Other carcinomas	5	2	7	8	4	10	8	4	11	6	9	6	7	5	6	4	8	1	8	6	5	7	7	4	9	3	160
Pancreatoblastoma				2	1		1			1	1	2					1				1				1	11
Pleuropulmonary blastoma							2	1	1		2		1	3	1		4		3		3	1	3	3			28
Other specified malignant				1				2	2			1	1	1		1				1							10
Unspecified malignant	2	1	5	3	2	4	4		1				1	1	1			1		1							27
Lymphoproliferative disease			1						1	4	3	3	3	4	4	3	3	4	3	4		1	3	1		3	48
LCH single system	10	15	20	22	27	26	30	33	47	23	30	38	27	28	32	25	37	30	24	33	30	34	39	4			664
LCH multi system	10	13	26	20	17	21	19	18	17	5	6	11	10	9	8	7	8	3	9	3	3	2	12	8	1		266
LCH unspecified	2		2			4	1			4	2			1			2		4	4	1	3	4	34	35	14	117
HLH	3	2		2	3	2	6	6	12	9	5	10	5	14	4	5	6	13	6	10	18	10	14	14	10	3	192
Ganglioneuroma	1	1	3	7	7	6	13	10	9	7	3	6	10	7	12	10	5	12	5	9	5	4	10	11	3	2	178
Other non-malignant peripheral nervous cell			3	1	2	3	5	2	3	3	4	6	5	2		2	2	4	2	2	2	3	1	4	5	2	68
Non-malignant embryonal renal	2	6	7	11	6	6	5	9	12	7	5	3	7	7	4	3	6	7	4	5	9	11	5	7	14	4	172
Non-malignant bone		1	1	1	5	3	3	3	4	2	2	1	1	5	1	3	1	5	2	4	1	2	4	6	7	2	70
Fibromatosis			1	4	3	4	6	7	10	3	6	10	4	5	7	5	6	10	5	5	4	3	10	9	10	6	143
NF & neurofibromatosis		3	4	3	4	13	8	9	11	11	6	5	11	12	7	8	7	12	7	2	9	10	12	6	15	5	200
Other non-malignant soft-tissue	3	3	8	4	12	9	8	13	22	8	6	11	22	14	19	11	13	11	15	20	20	30	33	23	43	8	389
Non-CNS non-gonadal non-malignant germ cell	5	10	17	8	13	10	13	17	27	16	16	7	14	15	18	13	18	13	19	10	13	24	18	23	16	3	376
Gonadal non-malignant germ-cell	1	1	6	4	7	6	17	12	13	12	15	14	13	15	16	18	11	20	14	12	17	13	21	14	15	2	309
Other non-malignant gonadal	1		2	2		2	3	5	7	4	8	4	4	8	5	6	4	7	5	4	6	8	7	7	12	4	125
Adrenocortical adenoma		2	2	2			1	2	2	2	3	3	1	3	1		3	2	2	4	2	2	2	2	1	2	46
Other non-malignant		1	3	2	4	4	6	5	7	3	12	7	16	6	12	10	10	4	8	11	6	10	12	16	8	9	192
Total	1087	1548	1795	1921	2088	2307	2476	2644	2957	1595	1557	1616	1641	1661	1593	1669	1822	1695	1726	1669	1697	1675	1835	1775	1700	724	46473

TABLE 1.5(i) CCLG REGISTRATIONS FOR CHILDREN AGED UNDER 15, BY CENTRE AND DIAGNOSTIC GROUP, 2001-2005

Centre	Leukaemia	Lymphomas	CNS	SNS	Retino-blastoma	Renal	Hepatic	Bone	Soft tissue sarcoma	Germ-cell etc	Epithelial	Other malignant	Other non-malignant	**Total**
Aberdeen	25	5	11	7		4		3	6	2	1		7	71
Barts/RLH	79	18	3	9	79	11	2	2	3	3	4		4	217
Belfast	56	21	37	9	2	20	3	7	12	3	1	1	6	178
Birmingham	244	64	164	40	79	41	11	25	45	25	17		70	825
Bristol	123	43	109	29	2	33	5	15	24	15	11	2	48	459
Cambridge	128	39	109	23	4	23	3	12	32	13	4	1	29	420
Cardiff	80	22	57	13	2	13	1	8	19	9	2	1	28	255
Dublin	184	65	87	50	7	39	11	32	46	11	4	2	44	582
Edinburgh	77	27	64	18	3	15		10	24	9	1		25	273
GOS	271	42	233	83		60	16	6	44	23	8	1	112	899
Glasgow	112	39	56	19	2	12	4	16	34	4	2	1	14	315
Leeds	144	61	100	24	4	19	5	20	23	15	8		54	477
Leicester	50	12	1	16	1	12		6	8	4		1	11	122
Liverpool	153	41	127	20	1	22	6	13	22	8	3		51	467
Manchester	182	52	136	32	3	40	6	30	37	17	13	1	65	614
Middlesex/UCLH	55	40	4	4		5	1	59	13	3	4		5	193
Newcastle	115	42	120	25	5	17	6	15	30	13	10		25	423
Nottingham	67	34	101	13		7	1	9	13	5	3	1	25	279
Oxford	101	25	92	24		22	1	11	21	6	6		18	327
Royal Marsden	152	73	141	36		40	9	16	26	18	10		13	534
Sheffield	81	26	69	15	11	18	4	14	13	12	7		24	294
Southampton	114	33	68	22		26	6	16	27	14	5		26	357
Total	2593	824	1889	531	205	499	101	345	522	232	124	12	704	8581

TABLE 1.5(ii) CCLG REGISTRATIONS FOR CHILDREN AGED UNDER 15, BY CENTRE AND DIAGNOSTIC GROUP, 2006-2011

Centre	Leukaemia	Lymphomas	CNS	SNS	Retino-blastoma	Renal	Hepatic	Bone	Soft tissue sarcoma	Germ-cell etc	Epithelial	Other malignant	Other non-malignant	**Total**
Aberdeen	16	4	10	7		4	1	2	4	2		1	4	55
Belfast	67	27	39	11	9	8	7	12	14	6	3		4	207
Birmingham	248	73	194	35	110	45	10	28	50	29	12	1	99	934
Bristol	153	42	123	40	5	27	6	28	43	20	9		51	547
Cambridge	179	55	125	35	7	28	3	13	39	29	9		106	628
Cardiff	100	19	79	15	1	14	3	9	25	6	8		34	313
Dublin	185	53	133	46	13	35	7	13	43	15	4	2	40	589
Edinburgh	70	10	70	12	1	13	1	7	9	3	3		33	232
GOS	276	65	281	67	9	76	11	2	54	29	3	1	140	1014
Glasgow	120	31	81	23	5	14	5	19	26	14	4		54	396
Leeds	127	51	102	38	2	22	8	23	29	23	6		51	482
Leicester	64	12	7	8	2	5		5	12		2		14	131
Liverpool	118	46	113	24	9	16	5	19	25	18	5	1	58	457
Manchester	168	68	133	33	5	28	7	27	47	18	11		77	622
Middlesex/UCLH	34	38	15	2		1		58	17	12	10		14	201
Newcastle	115	38	117	27	2	18	2	18	24	14	12		34	421
Nottingham	100	45	111	17	3	32	8	10	16	11	4	1	21	379
Oxford	114	35	99	20	7	13	2	10	12	12	4		38	366
Royal Marsden	241	90	163	41		48	10	15	41	19	15	3	33	719
Sheffield	100	39	69	15	3	19	4	11	15	5	4	1	37	322
Southampton	131	36	66	21	15	26	8	18	20	10	5	1	34	391
Total	2726	877	2130	537	208	492	108	347	565	295	133	12	976	9406

2. Patterns of Referral to CCLG Centres

The National Registry of Childhood Tumours receives copies of all notifications to cancer registries in the UK for children aged under 15 at diagnosis. By linking these records with CCLG (formerly UKCCSG) registrations the proportion of children initially referred to CCLG centres can be estimated as the proportion notified by cancer registration who have also been registered at diagnosis with the CCLG. Children with a CCLG registration who have not been notified through general (all ages) cancer registration cannot be included in these calculations as the number of non-CCLG patients who have also not been notified by cancer registration is unknown. Ascertainment by the Northern Ireland Cancer Registry is considered to be virtually complete only for 1993 onwards. Therefore, for continuity between calendar periods, the analyses of national data in Tables 2.1 and 2.2 refer to Great Britain only.

Analysis by diagnostic group and age at diagnosis in Great Britain

Table 2.1 shows the estimated proportions of children aged under 15 in the principal diagnostic groups who were registered with UKCCSG/CCLG during successive calendar periods. By 1998-2002, 90% of children with cancer were registered at diagnosis from a CCLG centre. Between the 1980s and 1990s there was a marked increase in percentage referred for CNS tumours and bone sarcomas. The only main diagnostic groups with a referral rate below 85% were epithelial tumours (two thirds of which were melanomas and carcinomas of the thyroid and skin) and the small and heterogeneous group of other and unspecified tumours. In 2008-10 the registration rate for retinoblastoma fell to 70% but this resulted from late registration for a time at one national referral centre rather than any decrease in the referral rate.

Table 2.2 shows the estimated proportions referred by age group during successive calendar periods. The referral rate has always been relatively low for children aged 10 years and above, and especially for those aged 13-14, but the gap has diminished.

Analysis by country and region of residence in the UK

Table 2.3 shows the estimated proportions of children referred by region of domicile. In England, the classification is by Strategic Health Authorities. In the most recent years, at least 80% of children in every English region, Wales and Scotland were referred to UKCCSG/CCLG centres. The referral rate in Northern Ireland was considerably lower until recently, largely because referral of children with CNS tumours was only 11% in 1993-97 and 24% in 1998-2002, but this proportion increased to 56% in 2003-10.

Langerhans cell histiocytosis

LCH is not routinely registered by cancer registries. A recent study from the BPSU, CCLG and Newcastle University estimated incidence per million child years in the UK and Ireland as 9.9 at age 0, 4.8 at age 1-4, 4.5 at age 5-9 and 1.8 at age 10-14 [Salotti et al., 2009]. Table 2.4 shows numbers of cases of LCH initially referred to a UKCCSG/CCLG centre in successive calendar periods with estimates of the referral rate based on expected numbers derived by applying the rates from this study to the child population of Great Britain. The estimated percentage of children with LCH who were referred to a UKCCSG/CCLG centre rose from 38% in 1978-82 to around 80% in 1998-2007 and 98% in 2008-10.

Referral by centre and region of residence

Table 2.5 shows the regions of residence for children registered at each centre during 1978-92, 1993-97, 1998-2002, 2003-2007 and 2008-2011. The great majority of children were treated at centres within their region of residence or an adjoining region. Referral patterns within South East England (London and South East Coast) have been complex but children have usually been referred to one of the London centres. In more recent years, a higher proportion of patients at GOS have been London residents and there has been a corresponding shift from GOS to Royal Marsden as the main referral centre for the South East Coast.

Referral by Cancer Network (England and Wales)

Since 2001, England has been covered by Cancer Networks, numbering 28 at the end of 2011, whose areas are each comprised of those of several Primary Care Trusts. A further three Cancer Networks cover Wales. Table 2.6 shows the CCLG centres from which children resident in each Cancer Network area were registered during 2001-2005 and 2006-2011. Retinoblastoma is excluded from this table because many children are registered from the two supraregional referral centres for this tumour. Thirteen of the 28 English Networks and all three Welsh Networks had more than 80% of their registrations from a single centre in both periods. More complicated referral patterns were seen in the remaining English Networks, predominantly in a large area of southern England including much of East Anglia, but also in the Severn valley and Humber and Yorkshire coast areas.

In 2006-2011 there were a number of changes in referral from 2001-2005, many of which were direct, or presumed indirect, consequences of the cessation of paediatric oncology at Barts/RLH. In 2001-2005, Barts/RLH accounted for 37% of referrals in NE London and 6-11% of those in North London and Essex. In 2006-2011, those components of referral seem largely to have moved to GOS in North East and North London and to UCLH in Essex. For NW London, GOS and UCLH both saw increases in registrations while registration of children from this Network ceased at Royal Marsden. The increase in referrals to GOS from the Networks mentioned above was offset by reductions in referrals from Anglia (with corresponding increase at Cambridge) and from SE London, Kent & Medway and Sussex (with increases mainly

at Royal Marsden). In Sussex, a reduction in referral to Southampton was also balanced by an increase at Royal Marsden.

Referral by Health Board (Scotland)

Table 2.7 shows CCLG centre of non-retinoblastoma registrations by Health Board for 2001-2011. As in the analysis of referral by Cancer Network, centres with fewer than 5% of the registrations for a Health Board are not shown. Most Health Boards had at least 80% of their registrations at a single centre. The exceptions were Central (Forth Valley), Highland and Orkney.

Irish Republic

Individual cancer registration records are not received from the Irish Republic. Table 2.8 shows estimated referral rates for 1994-2000 based on published data from the National Cancer Registry of Ireland (Stack *et al*, 2007). Referral rates were lower than in the UK overall and for several diagnostic groups, notably leukaemia, CNS tumours and retinoblastoma.

Table 2.9 shows CCLG registrations for 2001-2009. The annual number of registrations was 112 compared with 77 in 1994-2000. The size of the population at risk was similar during the two periods. The annual numbers of CCLG patients in 2001-2009 and cancer registrations in 1994-2000 were very similar for leukaemias, lymphomas, retinoblastoma and bone tumours. There were more CCLG registrations than cancer registrations for several diagnostic groups, notably neuroblastoma etc., renal tumours, hepatic tumours and soft tissue sarcomas, indicating increased incidence for these groups. For CNS tumours, germ-cell and gonadal tumours, epithelial tumours and other and unspecified tumours, the annual number of CCLG patients in 2001-2009 was lower than the annual number of cancer registrations in 1994-2000, suggesting that incidence has decreased or referral to CCLG was incomplete.

Table 2.1 Percentages of children aged under 15 with cancer or non-malignant CNS tumour initially referred to UKCCSG/CCLG, classified by diagnostic group. Great Britain 1978-2010

Diagnostic Group		Year of diagnosis						
		1978-82	1983-87	1988-92	1993-97	1998-2002	2003-2007	2008-2010
I	Leukaemia	69	81	86	92	94	97	96
	Lymphoid	70	83	88	92	95	98	97
	AML	71	81	87	98	97	95	93
	CML	43	67	68	69	75	95	100
	JMML/CMML	65	100	100	100	94	98	91
	Myelodysplasia	-	-	78	70	69	63	41
	Other & unspecified	43	32	38	66	79	80	72
II	Lymphomas	69	78	84	91	92	92	93
	Hodgkin lymphoma	64	72	77	89	93	96	95
	NHL & other lymphoma	74	83	88	92	92	88	90
III	CNS etc	32	41	56	81	89	89	87
	Choroid plexus tumours	16	44	59	88	89	95	90
	Ependymoma	30	47	63	90	93	93	93
	Astrocytoma	26	40	54	82	90	91	89
	Embryonal	49	56	71	94	98	96	97
	Other gliomas	31	39	55	81	92	93	90
	Craniopharyngioma	22	15	40	72	86	92	87
	Other CNS	26	21	27	56	62	71	64

- Fewer than 10 children with cancer registrations in this category

Table 2.1 (continued)

Diagnostic Group		Year of diagnosis						
		1978-82	**1983-87**	**1988-92**	**1993-97**	**1998-2002**	**2003-2007**	**2008-2010**
IV	Neuroblastoma etc	79	93	95	99	99	98	95
	Neuroblastoma	81	93	95	99	99	98	95
V	Retinoblastoma	10	68	84	94	92	92	70
VI	Renal tumours	75	91	93	97	98	99	98
	Wilms tumour etc	75	91	93	97	99	99	98
	Renal carcinoma	-	-	-	83	-	73	-
VII	Hepatic tumours	65	75	83	96	93	96	93
	Hepatoblastoma	71	83	91	98	97	100	94
	Hepatic carcinoma	45	53	63	-	71	88	-
VIII	Bone tumours	48	66	66	91	94	95	88
	Osteosarcoma	41	58	57	92	95	96	92
	Ewing sarcoma of bone	59	78	83	97	96	98	97
	All other malignant bone	32	45	25	50	63	50	19
IX	Soft tissue sarcomas	67	81	84	88	88	88	91
	Rhabdomyosarcoma	77	90	94	97	98	98	98
	Fibrosarcoma etc	51	49	61	63	74	68	76
	Extraosseous ESFT	85	88	91	96	99	97	95
	Synovial sarcoma	33	60	75	81	82	78	88
	All other specified	45	54	52	68	58	69	76

- Fewer than 10 children with cancer registrations in this category

Table 2.1 (continued)

Diagnostic Group		Year of diagnosis						
		1978-82	**1983-87**	**1988-92**	**1993-97**	**1998-2002**	**2003-2007**	**2008-2010**
X	Germ-cell and gonadal	49	63	71	84	87	87	90
	CNS germ-cell	39	36	63	88	85	96	91
	Other non-gonadal germ cell	69	94	82	85	92	86	85
	Malignant gonadal germ-cell	50	76	75	81	86	80	95
	All other malignant gonadal	-	21	-	-	-	-	-
XI	Epithelial	27	19	23	28	41	42	44
	Adrenocortical carcinoma	77	42	-	-	100	-	-
	Thyroid carcinoma	21	19	36	41	56	69	78
	Nasopharyngeal carcinoma	58	86	58	92	-	95	-
	Malignant melanoma	10	6	11	13	33	28	31
	Skin carcinoma	9	2	0	4	4	6	7
	Other carcinoma	29	22	29	35	40	40	43
XII	Other & unspecified	33	41	21	15	39	18	12
	Total	57	69	76	87	90	91	90

- Fewer than 10 children with cancer registrations in this category.

Table 2.2 Percentages of children with cancer or non-malignant CNS tumour initially referred to UKCCSG/CCLG, classified by age at diagnosis, Great Britain 1978-2010

	Year of diagnosis						
Age at diagnosis	**1978-82**	**1983-87**	**1988-92**	**1993-97**	**1998-2002**	**2003-2007**	**2008-2010**
0-9	62	74	81	90	93	93	91
10-12	54	63	68	81	88	86	87
13-14	35	45	51	70	79	84	83
Total	57	69	76	87	90	91	90

Table 2.3 Percentages of children in the UK with cancer or non-malignant CNS tumour initially referred to UKCCSG/CCLG, classified by country of domicile, and Strategic Health Authority (SHA) within England, 1978-2010

Region	1978-82	1983-87	1988-92	1993-97	1998-2002	2003-2007	2008-2010
North East	72	81	87	92	95	97	96
North West	78	79	83	91	95	94	93
Yorks & Humber	70	74	88	91	95	92	93
East Midlands	48	74	83	88	92	92	91
West Midlands	55	75	83	92	96	96	95
East of England	57	65	74	90	91	91	95
London	50	59	63	82	86	88	80
South East Coast	53	64	69	85	88	86	85
South Central	39	47	57	79	89	91	89
South West	52	64	78	86	87	89	91
England	58	69	76	88	91	90	90
Wales	60	64	77	76	84	86	96
England & Wales	58	68	76	87	91	91	90
Scotland	43	74	75	83	87	91	95
Great Britain	57	69	76	87	90	91	90
N Ireland				60	63	72	73
UK				86	90	90	90

Table 2.4 Expected numbers of incident cases of children with LCH and estimated percentage initially referred to UKCCSG/CCLG, Great Britain 1978-2010

Expected numbers are based on the incidence rates given by Salotti JA, Nanduri V, Pearce MS, Parker L, Lynn R, Windebank KP. Incidence and clinical features of Langerhans cell histiocytosis in the UK and Ireland. Arch Dis Child. 2009;94:376-80

	Year of diagnosis						
	1978-82	**1983-87**	**1988-92**	**1993-97**	**1998-2002**	**2003-2007**	**2008-2010**
Expected	220	210	217	221	215	208	128
UKCCSG/CCLG (%)	84 (38)	95 (45)	109 (50)	166 (75)	175 (81)	166 (80)	125 (98)

Table 2.5(i) Region of residence by centre for children in CCLG Register 1978-1992

Centre	North East	North West	Yorks/ Humber	East Midlands	West Midlands	East Anglia	London	South East Coast	South Central	South West	Wales	Scotland	Northern Ireland	Channel Islands	Isle of Man	Ireland	BFPO	Total
Aberdeen												148						148
Barts/RLH	8	39	24	29	14	201	240	106	79	27	19	8	12	2		7	5	820
Belfast		1							1				405					407
Birmingham	1	5		68	1536	3	2	1	5	5	18	1	1			1		1647
Bristol				1	3		1		7	960	4			2		1	3	982
Cambridge				16		423			4								1	444
Cardiff		1								1	526							528
Dublin																722		722
Edinburgh	2	1										335					2	340
Glasgow			1		1		1					789				2		794
GOS		2	2	40	2	582	770	460	156	34		1		10		1	30	2090
Leeds	8	6	987	4													1	1006
Leicester				221	1	1						1						224
Liverpool		565	1		3						145				22		2	738
Manchester		1279	6	27	2		1		1		4							1320
Middlesex/UCLH				1		7	15	5										28
Newcastle	753	104	12	1								3						873
Nottingham			6	469	1	1		1										478
Oxford				3	1				35	1								40
Royal Marsden		1	1	6	5	23	296	312	88	13	2	1		1		11	1	761
Sheffield			446	142								1					2	591
Southampton					1	1	1	16	364	131				17			4	535
Total	772	2004	1486	1028	1570	1242	1327	901	740	1172	718	1288	418	32	22	745	51	15516

Table 2.5(ii) Region of residence by centre for children in CCLG Register 1993-1997

Centre	North East	North West	Yorks/ Humber	East Midlands	West Midlands	East Anglia	London	South East Coast	South Central	South West	Wales	Scotland	Northern Ireland	Channel Islands	Isle of Man	Ireland	BFPO	Total
Aberdeen												68	1					69
Barts/RLH	4	27	11	16	6	78	151	51	17	14	10	3	5	1		1		395
Belfast													172					172
Birmingham		5	1	50	692		1			4	12		1					766
Bristol					5				3	522	3	1					2	536
Cambridge			1	9		253	1		2									266
Cardiff									1		223							224
Dublin																393		393
Edinburgh												200					1	201
Glasgow												320						320
GOS	1	1	5	5		263	471	181	25	7	2	1	2	4			4	972
Leeds	1	1	462														2	466
Leicester				98														98
Liverpool		297			4				1	1	68				5			376
Manchester		573	4	11	5						2							595
Middlesex/UCLH				1		35	69	21	2	2							1	131
Newcastle	324	25	2									2						353
Nottingham			1	238	6	2											1	248
Oxford				15	6	10			190	15								236
Royal Marsden						4	137	179	27	2	1					1		351
Sheffield			189	42														231
Southampton						1	1	18	211	84				7			4	326
Total	330	929	676	485	724	646	831	450	479	651	321	595	181	12	5	395	15	7725

Table 2.5(iii) Region of residence by centre for children in CCLG Register 1998-2002

Centre	North East	North West	Yorks/ Humber	East Midlands	West Midlands	East Anglia	London	South East Coast	South Central	South West	Wales	Scotland	Northern Ireland	Channel Islands	Isle of Man	Ireland	BFPO	Total
Aberdeen							1					70						71
Barts/RLH	4	9	8	8	3	52	176	23	14	13	4	7	3					324
Belfast													160					160
Birmingham	3	9	5	43	668	1	2	1	3	7	12	3	1					758
Bristol					3					472	1	1						477
Cambridge				11		364												375
Cardiff	1				1			1			285							288
Dublin																523		523
Edinburgh												252						252
Glasgow												292						292
GOS		1	3	1	3	221	450	141	14	7	3	2		9		1	1	857
Leeds	1	7	501														1	510
Leicester				106	5	2												113
Liverpool		328		1	4				1		82				7			423
Manchester		554	4	17	2	1	1	1		1								581
Middlesex/UCLH				5		48	104	21	8	1				1				188
Newcastle	351	44	8	1													1	405
Nottingham			3	306	16	2												327
Oxford				25	7	3	1		224	21								281
Royal Marsden				2		5	220	302	21	1								551
Sheffield		1	240	59														300
Southampton								49	191	83				4				327
Total	360	953	772	585	712	699	955	539	476	606	387	627	164	14	7	524	3	8383

Table 2.5(iv) Region of residence by centre for children in CCLG Register 2003-2007

Centre	North East	North West	Yorks/ Humber	East Midlands	West Midlands	East Anglia	London	South East Coast	South Central	South West	Wales	Scotland	Northern Ireland	Channel Islands	Isle of Man	Ireland	BFPO	Total
Aberdeen												62						62
Barts/RLH		4	2	1		14	54	8	4	3	1	1	1					93
Belfast							1						186					187
Birmingham	11	17	7	62	694	4	1		4	8	17	12				1		838
Bristol					6	1		1		458	1							467
Cambridge				14		455	2	2	2									475
Cardiff	1									2	260							263
Dublin						1										582		583
Edinburgh	1											236						237
Glasgow									1			311						312
GOS		2	2	3	1	209	581	122	11		1	2	2	1		3		940
Leeds	1	2	438	2		1											1	445
Leicester				121	7	3		1										132
Liverpool		340			6						86				13		1	446
Manchester		565		9	3						2							579
Middlesex/UCLH						42	126	21	4	1								194
Newcastle	349	40	7		1							1					1	399
Nottingham				271	17													288
Oxford				21	3	6	5		286	35							1	357
Royal Marsden						2	184	304	30									520
Sheffield	1		224	63	1	1			1									291
Southampton							1	23	212	104				11				351
Total	364	970	680	567	739	739	955	482	555	611	368	625	189	12	13	586	4	8459

Table 2.5(v) Region of residence by centre for children in CCLG Register 2008-2011

Centre	North East	North West	Yorks/ Humber	East Midlands	West Midlands	East Anglia	London	South East Coast	South Central	South West	Wales	Scotland	Northern Ireland	Channel Islands	Isle of Man	Ireland	BFPO	Total
Aberdeen												37						37
Barts/RLH																		
Belfast													135					135
Birmingham	4	17	20	41	499				4	4	14	6					1	610
Bristol										349	1							350
Cambridge				7	1	388	10	3										409
Cardiff								1		1	190							192
Dublin																354		354
Edinburgh										1		148						149
Glasgow												276						276
GOS		1	1	3	1	146	391	54	11	2	1	1	1	1				614
Leeds		2	293			1	1											297
Leicester				63	6	2	1											72
Liverpool		208		1	9						68	1			5	2		294
Manchester		393	3	7	2				1									406
Middlesex/UCLH						41	72	19	2									134
Newcastle	232	25	6									1						264
Nottingham			2	244	9	1												256
Oxford				7	1	7	6	1	176	28								226
Royal Marsden						7	185	291	16									499
Sheffield			153	39	1													193
Southampton							1	14	146	80				20				261
Total	236	646	478	412	529	593	667	383	356	465	274	470	136	21	5	356	1	6028

Table 2.6 CCLG registrations for all diagnoses except retinoblastoma by Cancer Network and CCLG centre, 2001-2005 and 2006-2011. For each combination of Network and period, the percentage of referrals is shown for all centres with at least 5% of that Network's registrations during the period.

Cancer Network	Centre	% of registrations	
		2001-2005	2006-2011
Lancashire & South Cumbria	Manchester	86	92
	Liverpool	10	7
Greater Manchester & Cheshire	Manchester	91	93
	Liverpool	9	7
Merseyside & Cheshire	Liverpool	99	98
North of England	Newcastle	97	98
Yorkshire	Leeds	98	98
Humber & Yorkshire Coast	Leeds	68	75
	Sheffield	31	24
North Trent	Sheffield	94	94
Greater Midlands	Birmingham	92	92
Pan Birmingham	Birmingham	98	99
Arden (Warwickshire)	Birmingham	92	93
East Midlands	Nottingham	53	62
	Leicester	22	21
	Birmingham	15	10
Anglia	Cambridge	74	84
	GOS	18	12
Essex	GOS	74	71
	UCLH	12	22
	Barts/RLH	11	
Mount Vernon (Beds, Herts)	GOS	52	43
	Cambridge	36	44
	UCLH	11	9
NW London	GOS	73	81
	UCLH	13	14
	R Marsden	8	
North London	GOS	72	85
	UCLH	23	11
	Barts/RLH	6	
NE London	GOS	50	81
	Barts/RLH	37	
	UCLH	12	16
SE London	GOS	50	29
	R Marsden	34	61
	UCLH	13	7
SW London	R Marsden	65	73
	GOS	30	22

Peninsula (Devon, Cornwall)	Bristol	98	98
Dorset	Southampton	96	97
Avon, Somerset & Wilts	Bristol	88	84
	Oxford	10	13
Three Counties (Herefs, Worcs, Gloucs)	Bristol	49	51
	Birmingham	47	43
Thames Valley	Oxford	91	88
Central South Coast	Southampton	93	96
Surrey, W Sussex & Hants	R Marsden	72	76
	GOS	18	18
	Southampton	5	
Sussex	R Marsden	67	82
	GOS	22	11
	Southampton	7	
Kent & Medway	R Marsden	51	68
	GOS	39	21
	UCLH	7	8
North Wales	Liverpool	97	100
Mid & West Wales	Cardiff	91	84
	Birmingham	8	12
SE Wales	Cardiff	99	98

Table 2.7 CCLG registrations for all diagnoses except retinoblastoma by Scottish Health Board and CCLG centre, 2001-2011. For each Health Board, the percentage of referrals is shown for all centres with at least 5% of that Board's registrations.

Health Board	Centres	% of registrations
Highland	Glasgow	75
	Edinburgh	14
	Aberdeen	11
Grampian	Aberdeen	90
	Edinburgh	6
Tayside	Edinburgh	98
Fife	Edinburgh	100
Lothian	Edinburgh	98
Borders	Edinburgh	93
Central (Forth Valley)	Glasgow	70
	Edinburgh	28
Argyll & Clyde	Glasgow	98
Greater Glasgow	Glasgow	100
Lanark	Glasgow	95
Ayrshire & Arran	Glasgow	100
Dumfries & Galloway	Glasgow	80
	Edinburgh	17
Orkney	Aberdeen	67
	Glasgow	17
	Edinburgh	17
Shetland	Aberdeen	83
	Edinburgh	17
Western Isles	Glasgow	100

Table 2.8 Childhood cancer in the Irish Republic 1994-2000

Total registrations are derived from Stack M, Walsh PM, Comber H, Ryan CA, O'Lorcain P. Childhood cancer in Ireland: a population-based study. Arch Dis Child 2007; 92:890-897

Diagnostic group	Total registrations	UKCCSG registrations	Estimated referral rate (%)
Leukaemia	237	156	66
Lymphoma	90	77	86
CNS tumours	215	119	55
Neuroblastoma etc	37	31	84
Retinoblastoma	16	6	38
Renal tumours	36	36	100
Hepatic tumours	4	3	75
Bone tumours	37	32	86
Soft-tissue sarcomas	50	45	90
Germ-cell & gonadal	27	20	74
Epithelial	27	11	41
Other & unspecified	11	2	18
Total	787	538	68

Table 2.9 Childhood cancer in the Irish Republic 2001-2009

Total numbers of children initially referred to a UKCCSG/CCLG centre 2001-2009, with 1994-2000 annual average UKCCSG referrals and total cancer registrations for comparison

Diagnostic group	UKCCSG/CCLG 2001-2009		UKCCSG 1994-2000	Cancer registrations 1994-2000
	Total	Annual	Annual	Annual
Leukaemia	338	37.5	22.3	33.9
Lymphoma	108	12.0	11.0	12.9
CNS tumours	201	22.3	17.0	30.7
Neuroblastoma etc	87	9.7	4.4	5.3
Retinoblastoma	20	2.2	0.9	2.3
Renal tumours	72	8.0	5.1	5.1
Hepatic tumours	18	2.0	0.4	0.6
Bone tumours	44	4.9	4.6	5.3
Soft-tissue sarcomas	80	8.9	6.4	7.1
Germ-cell & gonadal	27	3.0	2.9	3.9
Epithelial	7	0.8	1.6	3.9
Other & unspecified	4	0.4	0.3	1.6
Total	1006	111.8	76.9	112.4

3. Survival

Population-based survival 1971-2010

Table 3.1 and Figures 3.1-3.11 present population-based survival rates for all childhood cancers and for each of Groups I-X in ICCC-3. The results are for children from Great Britain who were diagnosed during 1971-2010 and included in the National Registry of Childhood Tumours. Most survivors have been followed up until the end of October 2012. Cases ascertained by death certificate only have been excluded.

Overall and for all diagnostic groups, the trend in survival by year of diagnosis was highly significant ($p<0.0001$). The largest percentage-point increase in five-year survival compared with the previous period for all cancers combined was in 1981-85. The largest such increases for leukaemias, lymphomas and neuroblastoma etc. were also in 1981-85. The largest increases were in 1976-80 for renal tumours, soft-tissue sarcomas and germ-cell and gonadal tumours, in 1986-90 for retinoblastoma and bone tumours and in 1991-95 for CNS tumours and hepatic tumours. The predominance of earlier calendar periods in these comparisons is quite largely accounted by the fact that as survival rates increased over time there was much less room for further large increases.

An alternative way of comparing changes in survival between successive periods is to calculate the reduction in the risk of death within 5 years from diagnosis as a percentage of the risk for the previous period. For all cancers combined the largest proportional reduction in the risk of death was in 1981-85 compared with 1976-80; the probability of death within five years for children diagnosed in 1981-85 was 0.392, a reduction of 22% from the probability of 0.504 in 1976-80. Among individual broad diagnostic groups, the largest percentage reductions in mortality were in 1976-80 for soft-tissue sarcomas (18%), in 1981-85 for lymphomas (37%), in 1986-90 for bone tumours (26%), in 1991-95 for CNS tumours (23%) and neuroblastoma etc (19%) and in 2001-05 for germ-cell and gonadal tumours (36%). For leukaemia, there were proportional reductions in mortality of 28% in 1981-85 and again in 2006-10. For retinoblastoma there were reductions in mortality of more than 50% in 1986-90, 2001-05 and 2006-10 but these were based on very small numbers of deaths. For renal tumours there was an alternating pattern of rising and falling survival rates from 1986-90 onwards; before then, the largest reduction in mortality was in 1976-80 (39%). For hepatic tumours, there were reductions in mortality of 49% in 1991-95 and 46% in 2006-10.

Survival of CCLG Patients 1978-2010

Tables 3.2-3.4 and Figures 3.12-3.132 present survival data for all CCLG patients registered at diagnosis throughout the UK and Ireland.

Table 3.2 gives five-year survival rates with the results of a test for trend for diagnostic groups with at least 50 registrations. For most of those with at least 150 registrations, survival rates are shown for five periods of diagnosis (1978-90, 1991-

95, 1996-2000, 2001-05, 2006-10) while for the remainder they are shown for two periods (1978-2000, 2001-2010). Table 3.3 gives overall five-year survival rates for selected smaller diagnostic groups and also for dysembryoplastic neuroepithelial tumour, for which there were more than 50 registrations but nearly all since 1996. Survival graphs are shown for all patients with a malignant neoplasm or non-malignant CNS tumour (Figure 3.12) and for each of the diagnostic groups in Tables 3.2 and 3.3 (Figures 3.13-3.41, 3.48-3.99). There were significant increases in survival rates for a wide range of diagnostic groups. In many instances, the greatest improvements took place in the 1980s but more recently there have been substantial improvements in survival for children with almost all types of leukaemia, NHL, ependymoma, medulloblastoma, neuroblastoma, Wilms tumour, hepatoblastoma, osteosarcoma, adrenocortical carcinoma, LCH and HLH.

Survival has also been analysed in relation to primary site for selected groups of CNS tumours (Table 3.4). Survival curves for children with astrocytoma, glioma or unspecified tumour in the brain stem are shown in figures 3.42-3.45. The survival rate for low-grade astrocytoma increased significantly over the study period but there was little evidence of improvement for high grade astrocytoma and other tumours in the same site. Figures 3.46 and 3.47 show survival curves for the two main types of spinal cord tumour, ependymoma and astrocytoma. Survival from spinal cord ependymoma increased significantly over time.

For selected diagnostic groups, survival rates have also been calculated for children diagnosed during calendar periods roughly corresponding to the periods of entry to successive trials. The results relate to all children diagnosed during a given period, not just those who were actually entered. The survival graphs for these are shown in Figures 3.100-3.132. In the keys to these graphs, calendar periods with no trial open are shown in brackets.

Results for ALL are presented separately according to Down syndrome (DS) status and age at diagnosis. Infants aged under one year, who have a markedly worse prognosis than older children, were excluded from some UKALL trials and since 1992 have had their own study protocols. DS is an adverse prognostic factor in ALL, and children with DS have sometimes been less likely to be entered in national trials.

Among non-DS children with precursor-cell ALL, survival of infants remained fairly constant throughout 1978-88. Survival increased successively thereafter and in 1999 onwards, the era of the INTERFANT 99 trial, five-year survival exceeded 50% for the first time.

Survival of older non-DS children with ALL increased with each successive trial period. The increase in survival was rather small between 1991-96 and 1997-99 (eras of UKALL XI and ALL 97), but this was followed by a larger increase in 2000-02 (era of ALL 97/99). One-year survival reached 96% in the era of UKALL XI and has remained at that level; the more recent increases in overall survival since then are the result of continuing decreases in subsequent mortality among one-year survivors. In 2003 onwards, the era of ALL2003, five-year survival was over 90%.

Children with DS and ALL have also experienced a substantial increase in survival rates. There was an especially large improvement in 1991-96 (era of UKALL XI) compared with previous years, when the gap between DS and non-DS children was greatly narrowed. In 1997-2002 (era of ALL97 and ALL 97/99), the survival rate decreased again. This was entirely due to a substantial fall in one-year survival from 92% in 1991-96 to 68% in 1997-2002. The increase in subsequent survival among patients who had survived one year that was achieved in the era of UKALL XI was maintained. In 2003-10 (era of ALL 2003) one-year survival increased to 85% and five-year survival was 69%.

At the start of the study period, the outlook for children with mature B-cell leukaemia was very poor, with fewer than a quarter surviving five years. Between the periods of the second and third series of NHL studies (1985-89 and 1990-95), five-year survival increased dramatically from 40% to 69%. Five-year survival since 1996 has been above 75%.

For AML, survival has also been analysed separately for children with and without DS. Among non-DS children, survival increased steadily between 1983 and 2004. During 2005-2008 (era of AML15), five year survival was 67%. Survival of DS children has also increased, but for this group there was no change before 1988. During 1988-94 (era of AML 10), survival was still lower than for non-DS patients, but during 1995-2002 (AML 12), DS children had the better prognosis, with five-year survival of 74% compared with 61% for non-DS children. This improvement was maintained during 2003-2010, with five-year survival reaching 83%. From the mid 1990s onwards, DS children with AML have had a markedly better outcome than those with ALL.

Five-year survival for Hodgkin lymphoma was already over 90% at the start of the first UKCCSG study in 1982; there have only been small increases since then. Separate protocols for T-cell and B-cell NHL were used throughout the study period. Survival for T-NHL rose between the eras of the first and second series of NHL studies but then showed no consistent pattern until 2004-2010 (era of the 2004/08 trial and subsequent non-trial period), when five-year survival was over 85% compared with 72-80% for children diagnosed throughout the previous 19 years. Survival from B-NHL increased with each successive trial period. Survival from anaplastic large cell lymphoma has been significantly higher since 1998, when specific trials for this subtype of NHL began.

Young age at diagnosis is an adverse factor for ependymoma. During 1992-2005, children under 3 years of age were eligible for the infant brain tumour study, and 5-year survival in that period was 54%. Survival of older children with ependymoma has also increased, exceeding 70% during 1999 onwards.

Survival from low-grade astrocytoma was higher during the era of LGG-1 (1997-2003) than previously, 93% compared with 88%. The small further increase since then was non-significant.

Young age at diagnosis is also an adverse factor for medulloblastoma and other PNET, and in the 1990s special protocols were available for the treatment of children aged under 3 years. Before then, some younger children – though no infants aged under a year – were included in trials where most patients were older. There has been no straightforward trend in survival for the 0-2 year age group. For older children, survival was lower in the mid to late 1980s than it was in the era of the first UKCCSG Brain Tumour Study. Five-year survival in 1992-2000 (era of the international PNET-3 trial) was 62%; it increased further, to 64%, during 2001-10, which included the era of PNET-4.

Age is also an important prognostic factor in neuroblastoma, with infants aged under a year having a much higher survival rate than children aged 1 year and over, and entry to most trials has been limited to one or the other of these age groups. Survival of infants in the era of ENSG8 (1992-98) was hardly different from earlier years. Five-year survival rose to 90% during 1999-2004 (1999 03 study era), followed by a decrease to 83% during 2005-10. For children aged 1-14 years survival in the period of ENSG1 and 3 (1982-1989) was higher than previously. Survival increased further since then and five-year survival has reached 55% in the era of the current high-risk neuroblastoma trial.

Survival from Wilms' tumour showed a steady increase between successive trial eras. In the era of the most recent SIOP trial (2002 onwards) the risk of death within one year from diagnosis was 3.1%, a better than one-third reduction from the one-year mortality rate of 5.1% in the preceding era (UKW-3 trial, 1992-2001). There is no evidence of any increase in survival from rhabdoid renal tumour or renal clear cell sarcoma during the entire study period.

Survival rates for hepatoblastoma during SIOPEL-1 (1990-94) were substantially higher than before but there was no further increase in the period of SIOPEL-2 (1995-97). Survival has increased again since the opening of SIOPEL-3 in 1998, with five year survival reaching 82%. There was no sign of a trend in survival rates for hepatic carcinoma.

For osteosarcoma, five-year survival during the MRC trial era ending in 1982 was 39%. Throughout the period 1983-2004, five-year survival fluctuated between 54% and 60%. There was a further increase to 67% in the era of the EURAMOS-1 trial from 2005 onwards. Survival rates for Ewing sarcoma of bone were markedly higher than previously during the era of the second UKCCSG study (1987-92) but have shown no change since then. For rhabdomyosarcoma there was little sign of a trend during 1978-88, with five-year survival of 53-57%. Survival throughout 1989-2005 (including the eras of MMT-89 and MMT-95) was 64-68%. In 2006 onwards (era of RMS 2005) there was a further small increase in 5-year survival to 71%.

There was a major change in the chemotherapy protocol for germ-cell tumours in 1983, part way through the period of entry to the first UKCCSG study. Survival rates for gonadal and other extracranial tumours increased sharply at the time of this change. Since the start of the second study in 1989 there has been a further improvement in survival for ovarian and extragonadal tumours. Survival from

intracranial germinoma and other types of CNS germ-cell tumours increased substantially to just over 90% since the opening of the SIOP study in 1997.

Five-year survival from single-system Langerhans cell histiocytosis (LCH) was already 95% during 1978-90, before the first LCH trial opened, but has increased still further since then. There was little change in survival from multi-system LCH from 1978-90 until the era of the third trial, starting in 2002, since when five-year survival has been above 95%. For haemophagocytic lymphohistiocytosis (HLH) five-year survival increased from 14% during 1978-94, before the start of HLH-94, to 34% during the era of HLH-94 (1995-2003). Survival increased further since then, to 65% during 2006 onwards.

The reader is also referred to the recent paper on population-based survival by eras of entry to clinical trials (Stiller, *et al,* 2012; full reference in section 4 below).

Table 3.1 Population-based survival of children with cancer in Great Britain diagnosed 1971-2010 by period of diagnosis

	Five-year actuarial survival (%)								
Diagnostic group	**1971-1975**	**1976-1980**	**1981-1985**	**1986-1990**	**1991-1995**	**1996-2000**	**2001-2005**	**2006-2010**	**X^2 (1df) for trend**
Leukaemias	33	45	61	67	75	79	83	88	2951***
Lymphomas	47	59	74	80	85	86	88	91	676.7***
CNS tumours	41	47	54	57	67	71	72	75	875.7***
Neuroblastoma etc	16	28	42	40	52	60	64	67	579.9***
Retinoblastoma	88	88	89	95	95	97	99	100	52.6***
Renal tumours	59	75	77	84	78	88	84	87	140.4***
Hepatic tumours	14	19	33	31	65	66	67	82	117.5***
Bone tumours	29	32	42	57	61	64	61	68	236.6***
Soft tissue sarcomas	37	49	58	61	64	65	68	71	168.9***
Germ cell & gonadal tumours	45	59	73	81	85	87	91	93	219.1***
All cancers	40	50	61	66	73	76	79	82	538.7***

***$p<0.001$

Table 3.2 Survival of CCLG patients diagnosed 1978-2010 by period of diagnosis. In the test for trend by year of diagnosis, brackets around the X^2 value indicate a negative trend. ***Results for certain diagnostic subgroups, mainly 'other and unspecified', whose composition may have changed over the years are printed in italics***.

	Five-year actuarial survival (%)							
Diagnostic Group	**1978-90**	**1991-95**	**1996-2000**	**2001-05**	**2006-10**	**1978-2000**	**2001-2010**	**X^2 (1df) for trend**
Precursor ALL	69	81	83	88	92			683.4***
Mature B-cell leukaemia						54	79	31.4***
AML	33	53	67	65	69			244.6***
CML	41	56	74	79	93			44.7***
Myelodysplasia	*21*	*55*	*53*	*66*	*63*			10.1**
JMML & CMML	*12*	*35*	*34*	*63*	*76*			*45.9****
Other & unspecified leukaemia						*46*	*66*	*21.6****
Hodgkin lymphoma	91	96	93	95	96			13.1***
NHL	68	78	81	86	88			106.7***
Ependymoma	42	65	66	65	71			23.7***
Choroid plexus papilloma						87	98	21.6***
Choroid plexus carcinoma						22	24	(0.05)
Low grade astrocytoma	84	90	92	94	95			60.0***
High grade astrocytoma	23	18	18	11	17			(1.79)
Unspec. Astrocytoma	*55*	*42*	*45*	*72*	*66*			*4.58**
Medulloblastoma	51	54	66	65	64			24.4***
CNS PNET	36	26	33	37	36			2.05
ATRT						18	24	0.13
Oligodendroglioma						64	62	(0.46)
Other glioma	25	28	41	38	46			19.8***
Pituitary carcinoma & adenoma						93	100	1.57

Table 3.2 (continued)

Diagnostic Group	1978-90	1991-95	1996-2000	2001-05	2006-10	1978-2000	2001-2010	X^2 (1df) for trend
Craniopharyngioma	91	90	99	94	97			3.73
Pineoblastoma						28	46	9.16**
Ganglioglioma						85	92	3.32
Meningioma						78	97	12.9***
Unspecified CNS	*36*	*65*	*55*	*32*	*49*			*0.06*
Neuroblastoma	40	52	59	64	65			174.7***
Retinoblastoma bilateral	92	93	98	99	100			19.6***
Retinoblastoma unilateral	92	97	96	98	99			8.83**
Wilms tumour	83	80	92	90	90			38.6***
Rhabdoid renal tumour						26	11	(1.71)
Renal clear cell sarcoma						78	85	0.01
Hepatoblastoma	40	71	79	77	86			46.8***
Hepatic carcinoma						23	29	0.01
Osteosarcoma	49	53	62	56	65			14.1***
Ewing sarcoma of bone	48	67	62	64	68			29.2***
Rhabdomyosarcoma	57	65	68	65	71			25.4***
MPNST						34	59	2.13
Other fibrosarcoma etc						72	90	11.1***
Extraosseous ESFT	51	53	55	66	58			3.46
Extrarenal rhabdoid tumour						15	21	0.10
Synovial sarcoma						81	81	1.31
Other specified soft tissue sarcoma						*62*	*66*	*0.08*
Unspecified soft-tissue sarcoma	*40*	*40*	*33*	*45*	*68*			*2.26*
Hepatic sarcoma, all types						48	56	0.67
Intracranial & intra-spinal germinoma	73	83	87	92	92			8.72**

Table 3.2 (continued)

Diagnostic Group	1978-90	1991-95	1996-2000	2001-05	2006-10	1978-2000	2001-2010	X^2 (1df) for trend
Other CNS germ-cell						50	72	10.2***
Other malig. extra-gonadal germ-cell	68	81	82	90	91			33.1***
Gonadal malig. germ-cell	92	96	96	97	99			21.0***
Adrenocortical carcinoma						36	63	16.1***
Thyroid carcinoma, non-medullary						100	99	(0.02)
Nasopharyngeal carcinoma						73	88	6.09*
Malignant melanoma						52	59	2.67
Misc. other carcinoma						*50*	*63*	*1.63*
LCH single system	95	97	99	99	99			9.01**
LCH multi system	68	72	73	97	96			11.4***
HLH	6	19	33	44	65			32.9***
Ganglioneuroma	100	100	97	100	100			0.25
Mesoblastic nephroma						99	96	0.04
Fibromatosis						94	98	3.66
Misc non-malig soft tissue	*95*	*97*	*97*	*100*	*99*			*2.06*
Other non-gonadal non-malig. germ-cell	96	98	97	99	100			1.76
Gonadal non-malig. germ-cell	100	100	99	100	100			0.15
Non-malignant specialised gonadal						94	98	0.03

* P<0.05
** P<0.01
*** P<0.001

Table 3.3 Survival of CCLG patients diagnosed 1978-2010

Diagnostic group	Five-year actuarial survival (%)
Pinealoma & Pineocytoma	79
Desmoplastic infantile astrocytoma	93
DNET	99
Renal PNET	28
Renal carcinoma	68
Chondrosarcoma	50
Leiomyosarcoma	84
Fibrohistiocytic tumours	92
Alveolar soft part sarcoma	95
Desmoplastic small round cell tumour	25
Other malignant gonadal tumours	53
Thyroid carcinoma, medullary	95
Salivary gland carcinoma	94
Colorectal carcinoma	16
Pleuropulmonary blastoma	78
Lymphoproliferative disease	66
Adrenocortical adenoma	100

Table 3.4 Survival of CCLG patients diagnosed 1978-2010 with selected CNS tumours. Note that these results relate to patients also included in Table 3.2.

	Five-year actuarial survival (%)							
Diagnostic group	**1978-90**	**1991-95**	**1996-2000**	**2001-05**	**2006-10**	**1978-2000**	**2001-10**	**X^2 (1df) for trend**
(i) Astrocytoma, glioma or unspecified tumour of brain stem								
Total	17	21	32	29	29			8.55**
Low-grade astrocytoma	39	59	74	80	79			16.8***
High-grade astrocytoma						5	3	0.10
Unspec astrocytoma, other glioma & unspecified	*13*	*13*	*26*	*18*	*21*			*2.30*
(ii) Spinal cord								
Ependymoma						93	100	8.54**
Astrocytoma	66	75	85	67	87			1.63

** P<0.01
*** P<0.001

Survival graphs

Figures 3.1-3.11 Population-based survival of children with cancer in Great Britain diagnosed 1971-2010 by 5-year period of diagnosis, 1971-75, 1976-80, … , 2001-05, 2006-10.
Source: National Registry of Childhood Tumours

Figures 3.12-3.99 Survival of CCLG patients diagnosed 1978-2010 by period of diagnosis, subdivided as 1978-90, 1991-95, 1996-2000, 2001-2005, 2006-2010 or 1978-2000, 2001-10 for diagnostic groups and subgroups with larger numbers of patients.
Source: National Registry of Childhood Tumours / CCLG

Figures 3.100-3.132 Survival of CCLG patients in selected diagnostic subgroups diagnosed 1978-2010 by period of diagnosis, subdivided into periods roughly corresponding to the periods of entry to successive trials. The results relate to all children diagnosed during a given period, not just those who were actually entered. In the keys to these graphs, calendar periods with no trial open are shown in brackets.
Source: National Registry of Childhood Tumours / CCLG

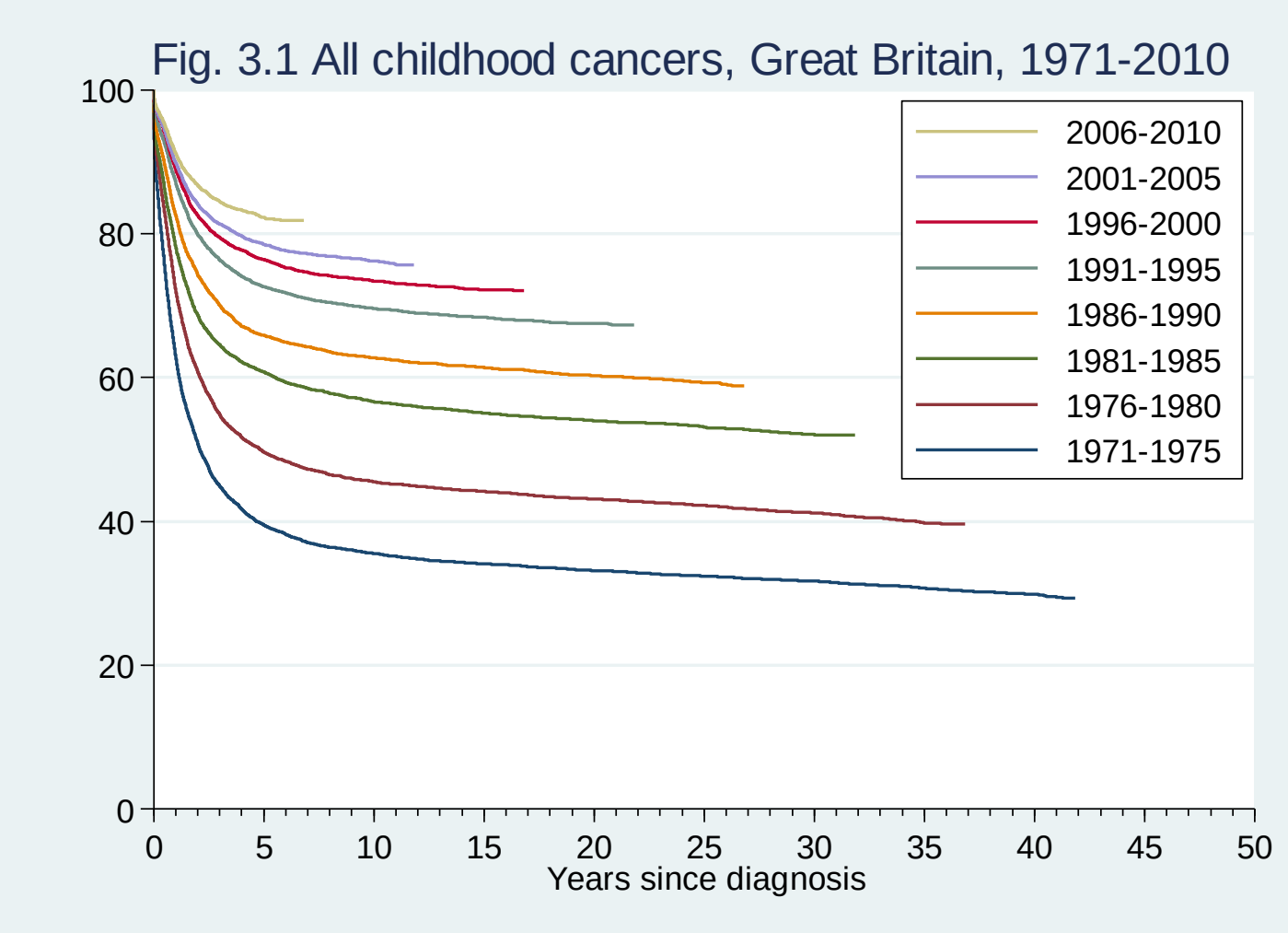


Fig. 3.2 Leukaemias, Great Britain, 1971-2010

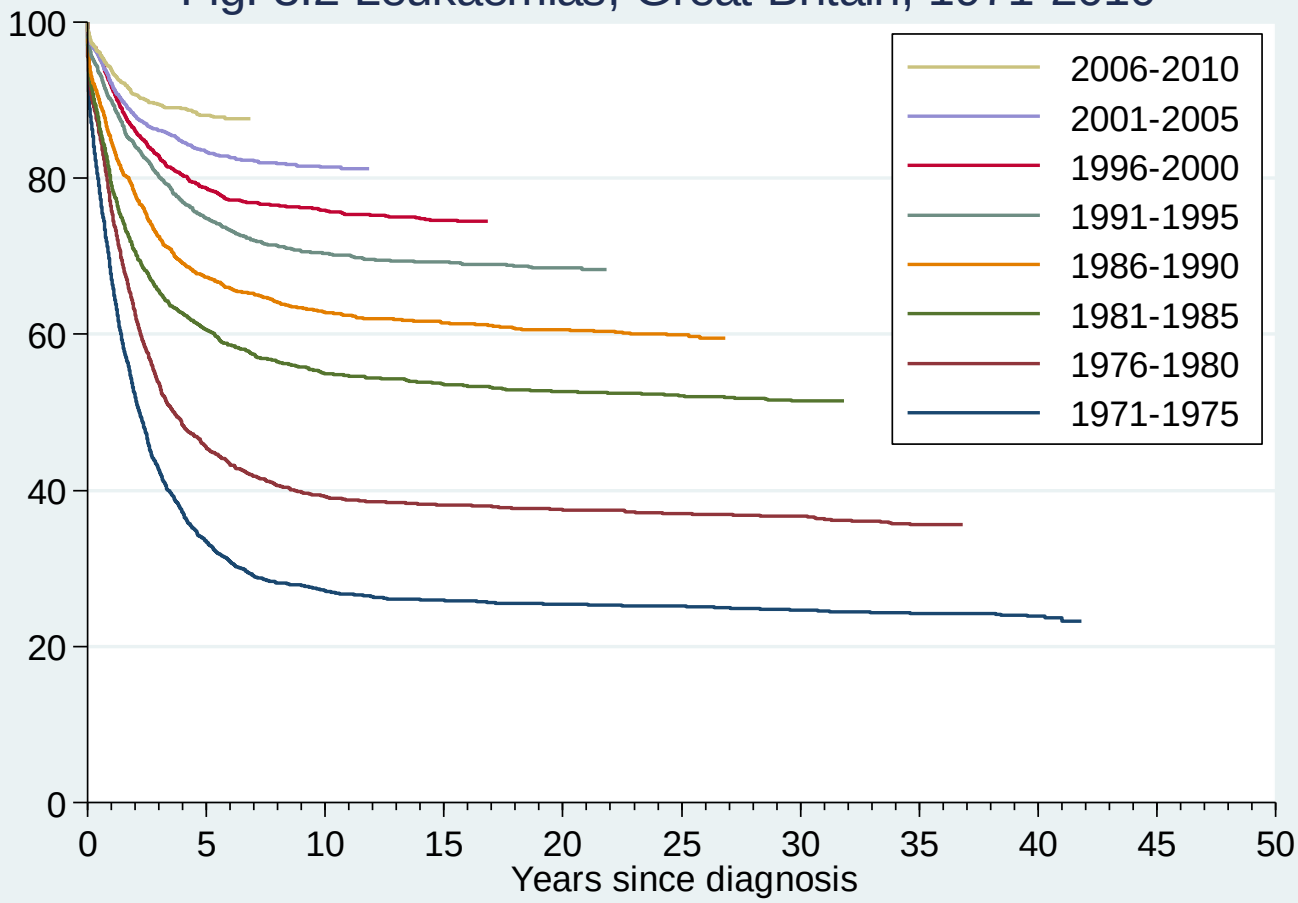


Fig. 3.3 Lymphomas, Great Britain, 1971-2010

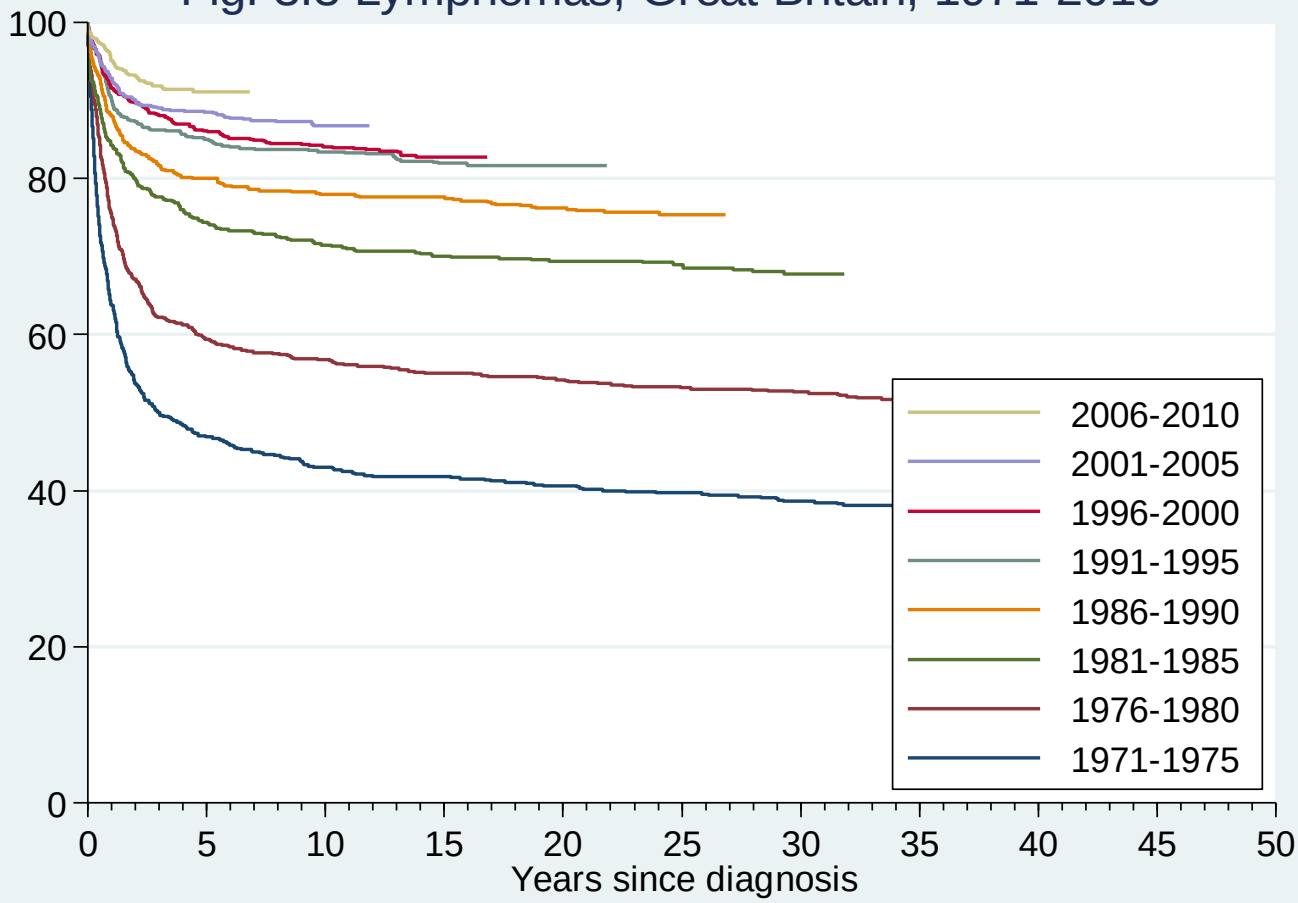


Fig. 3.4 CNS tumours, Great Britain, 1971-2010

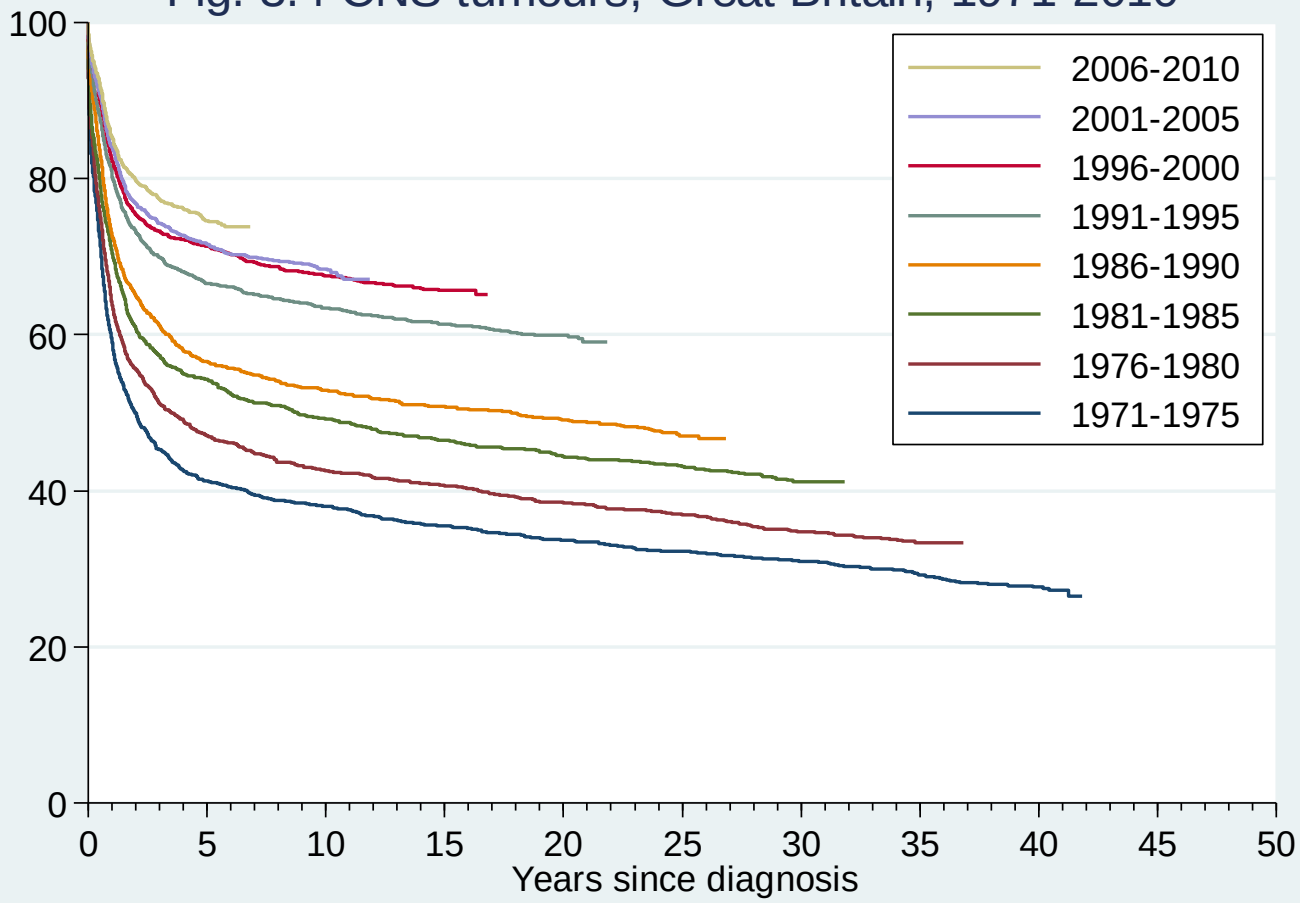


Fig. 3.5 Neuroblastoma etc., Great Britain, 1971-2010

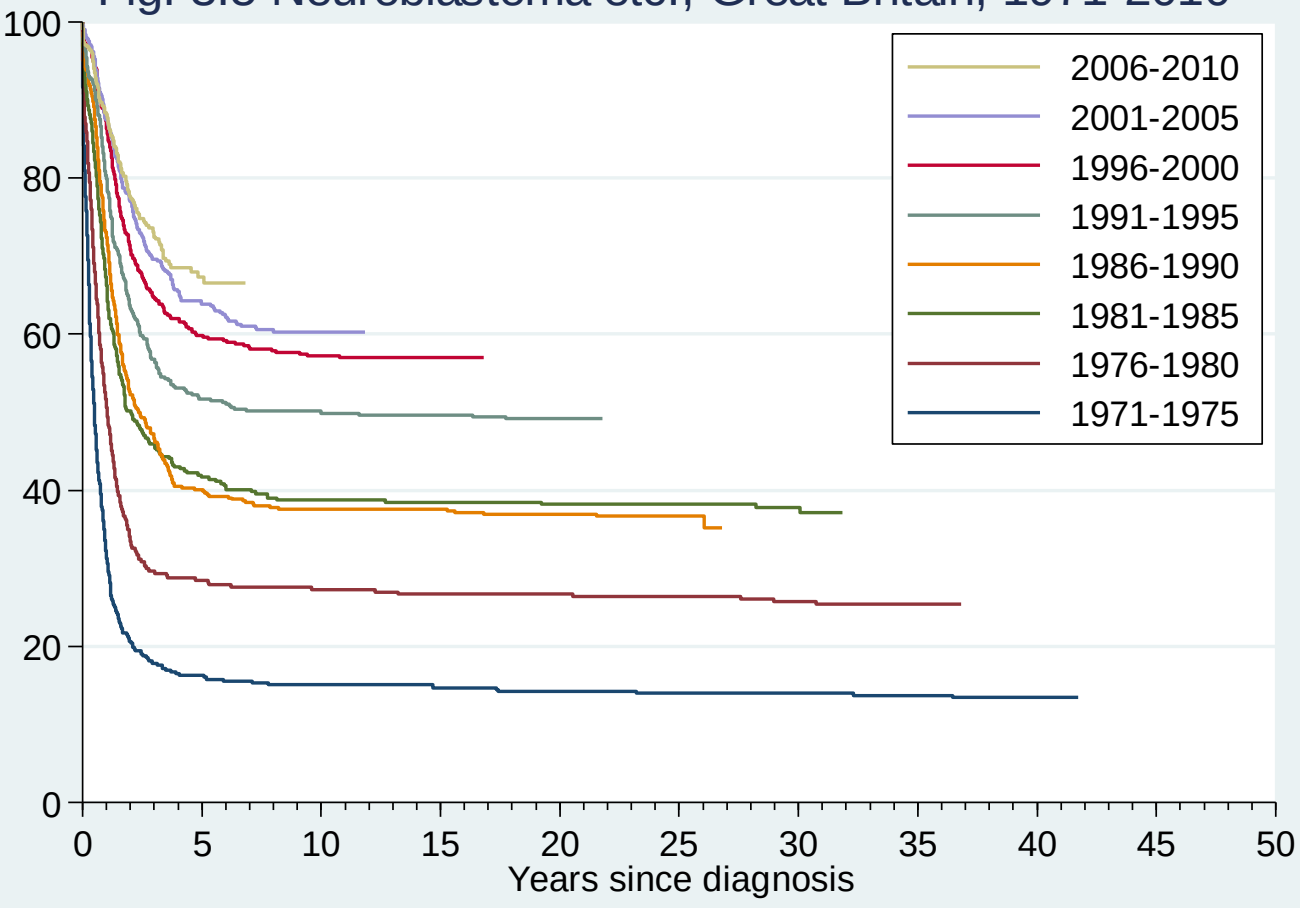


Fig. 3.6 Retinoblastoma, Great Britain, 1971-2010

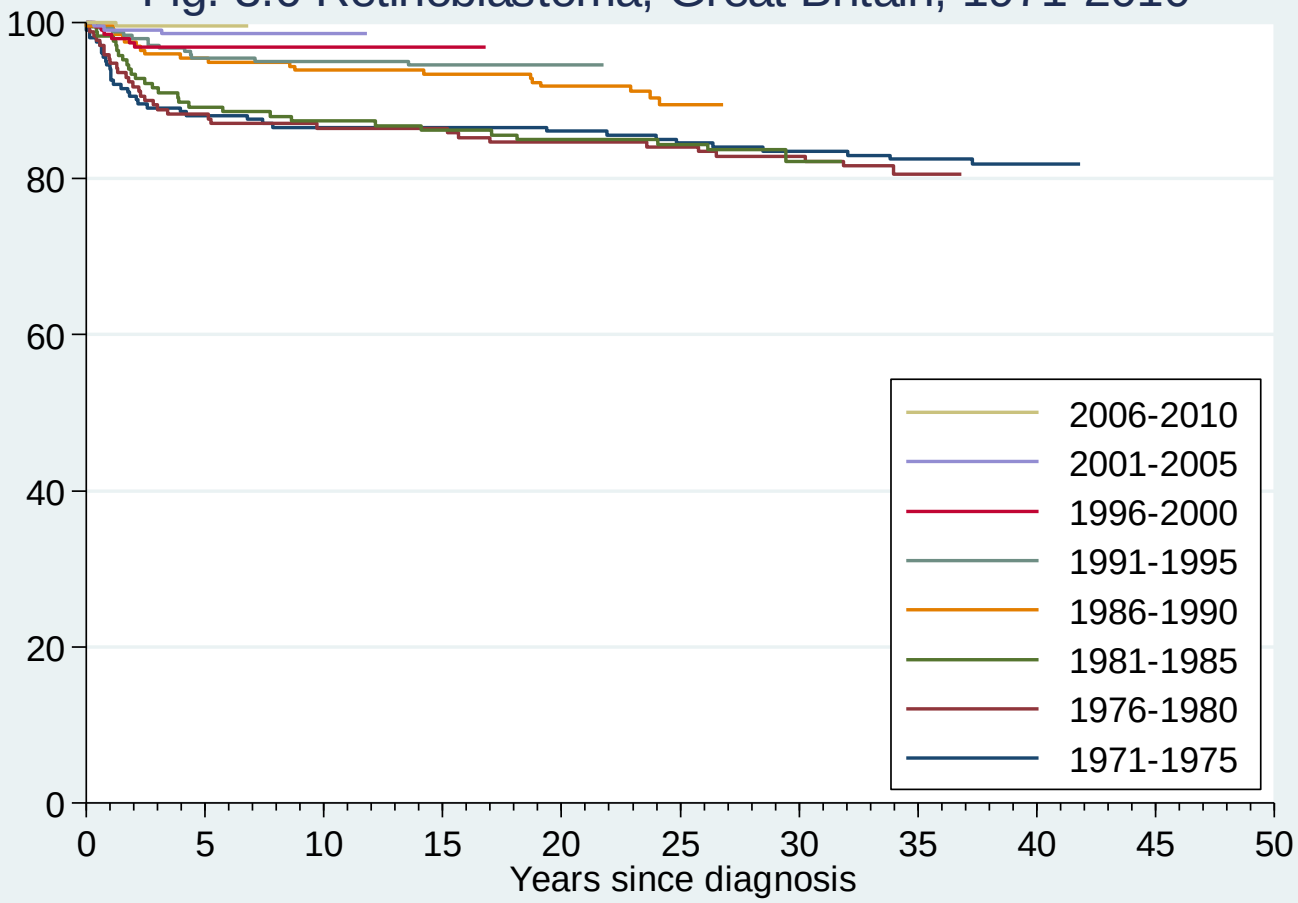


Fig. 3.7 Renal tumours, Great Britain, 1971-2010

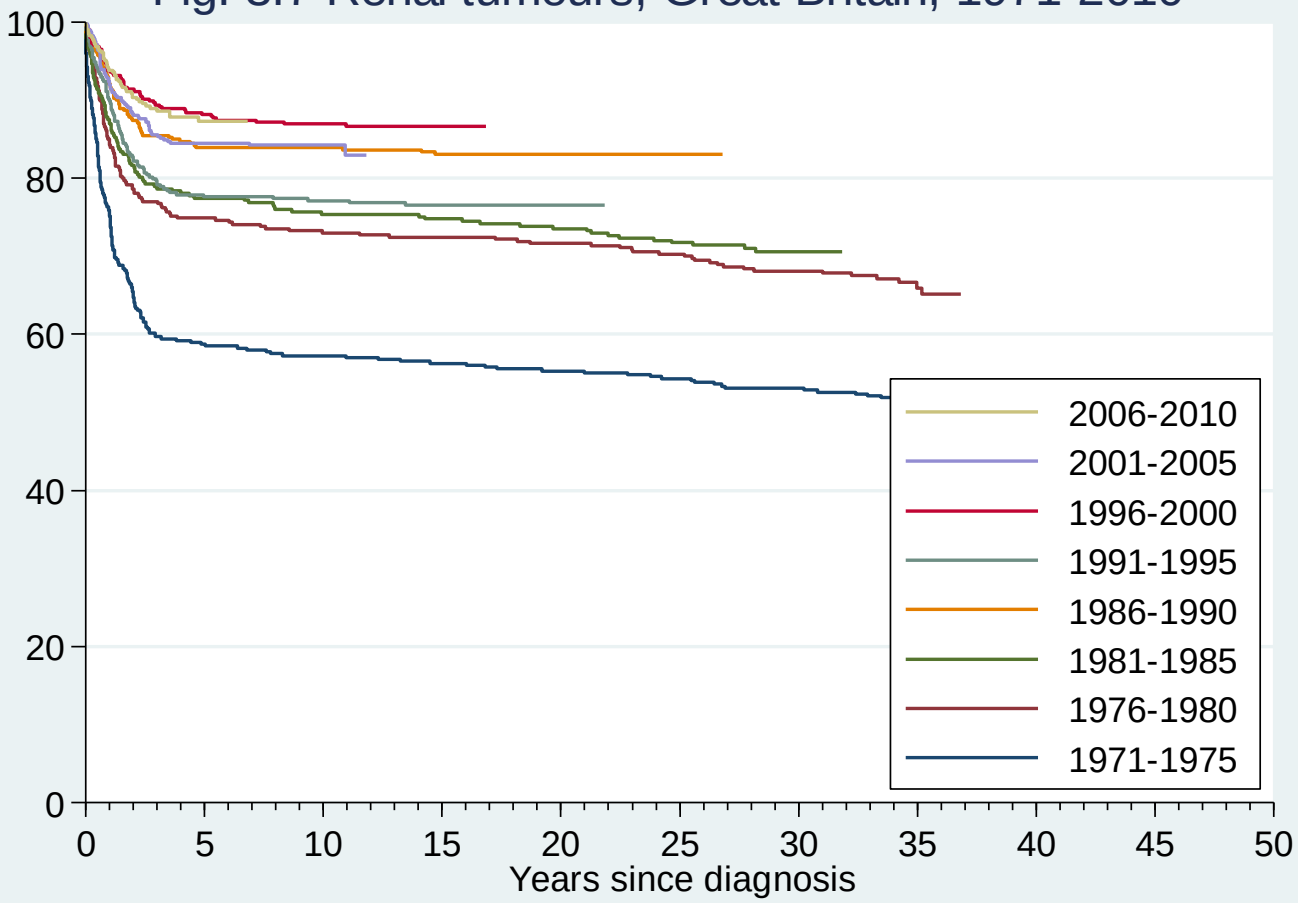


Fig. 3.8 Hepatic tumours, Great Britain, 1971-2010

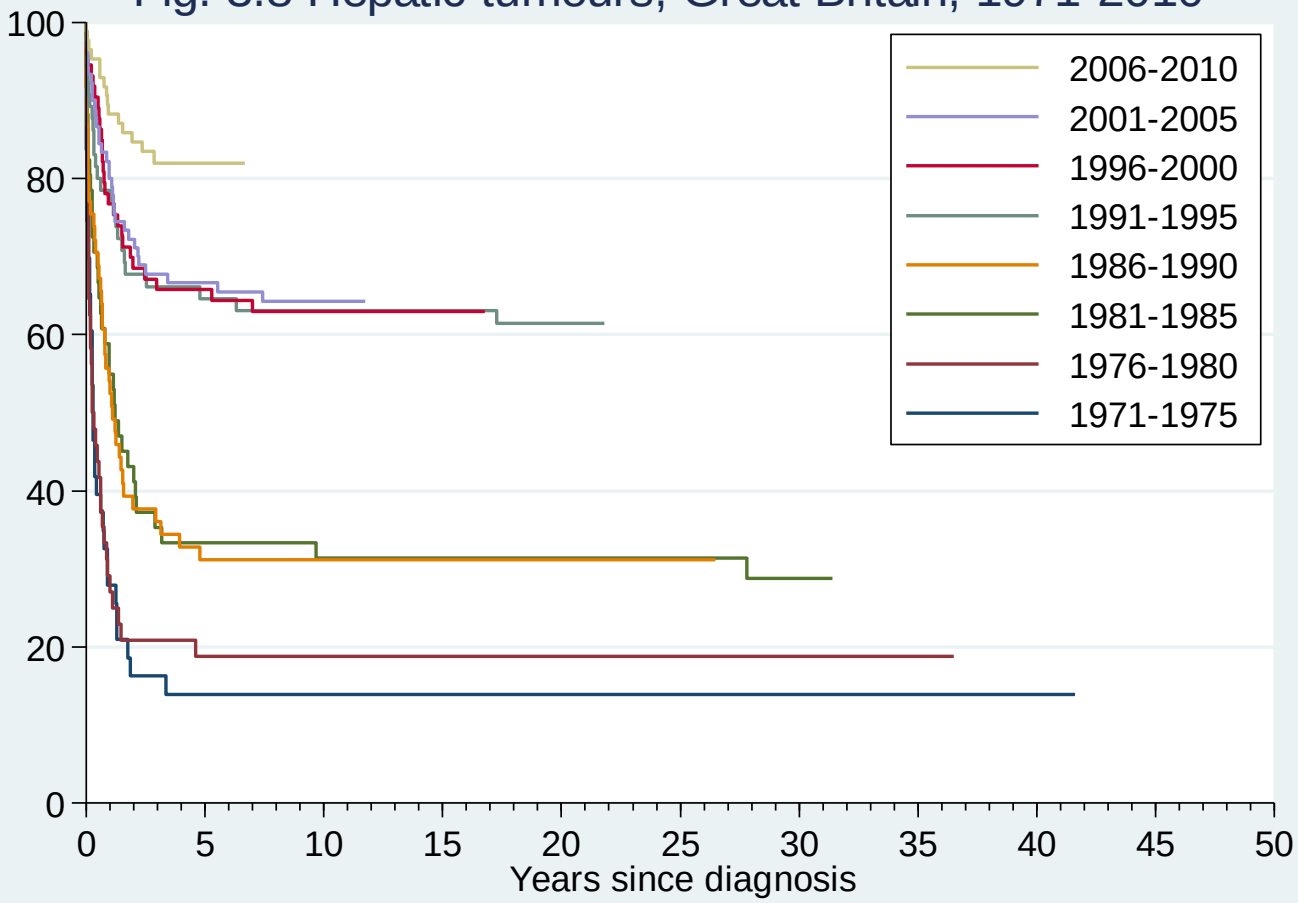


Fig. 3.9 Bone tumours, Great Britain, 1971-2010

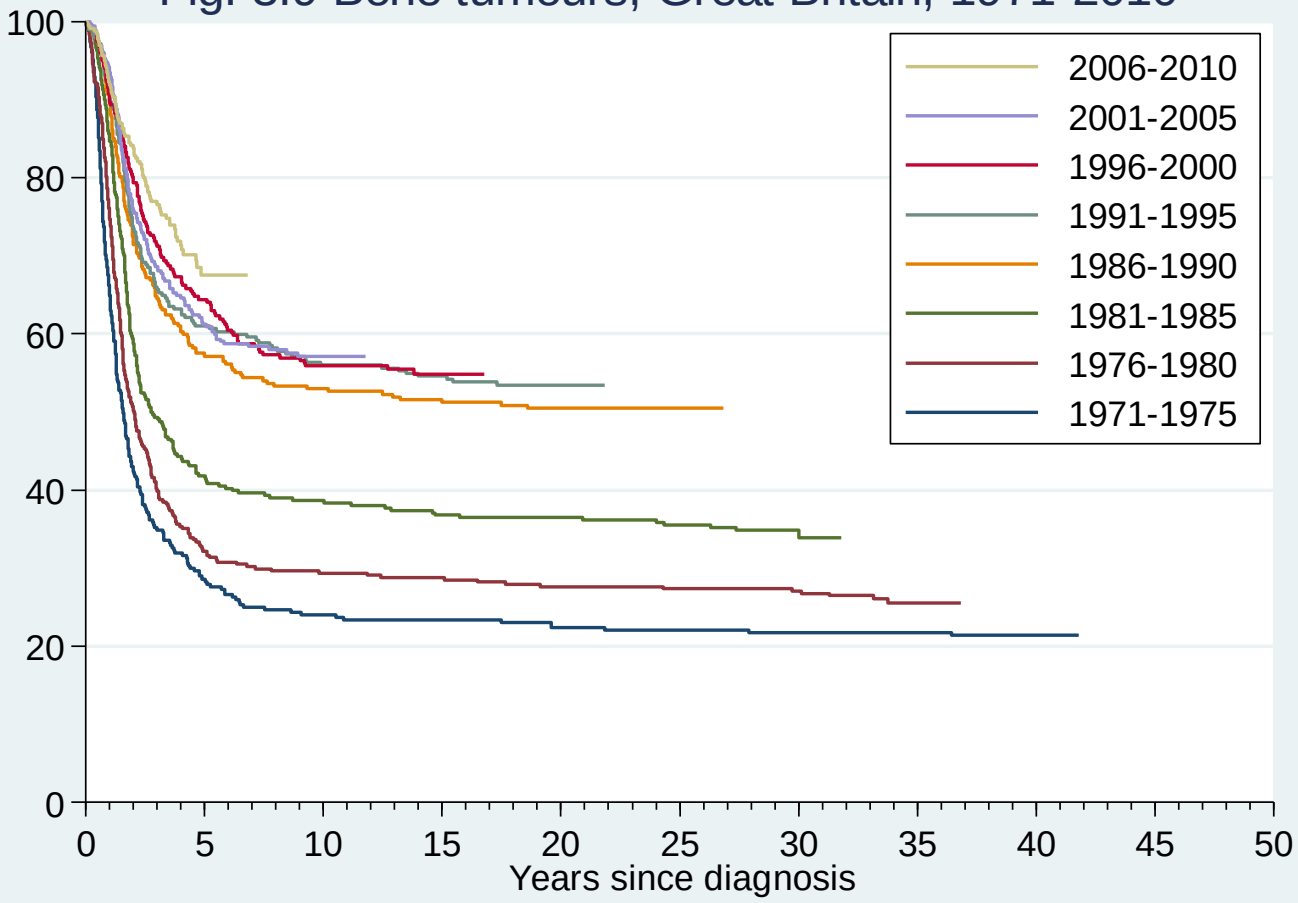


Fig. 3.10 Soft tissue sarcomas, Great Britain, 1971-2010

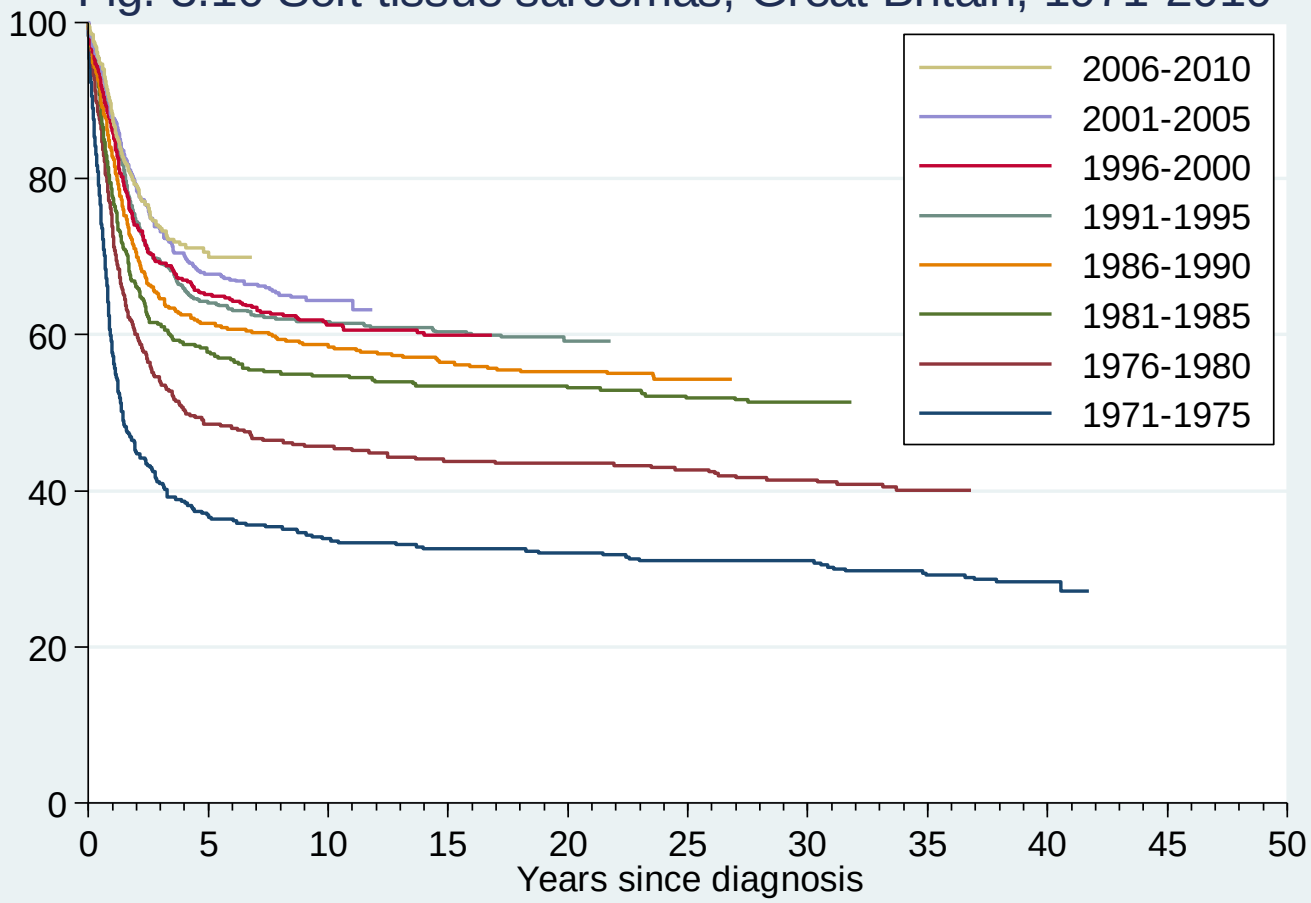


Fig. 3.11 Germ-cell & gonadal, Great Britain, 1971-2010

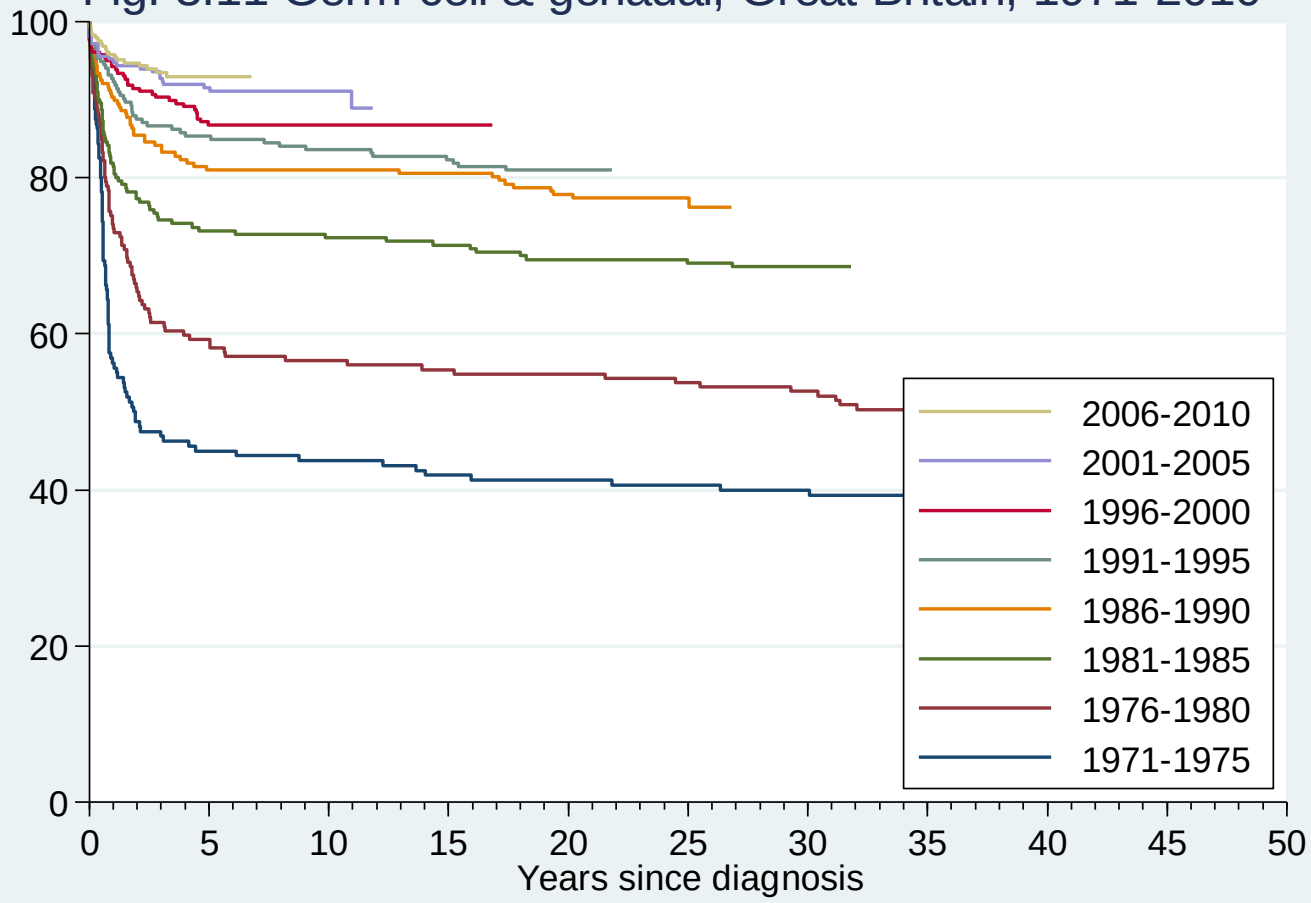


Fig. 3.12 All childhood cancers, CCLG

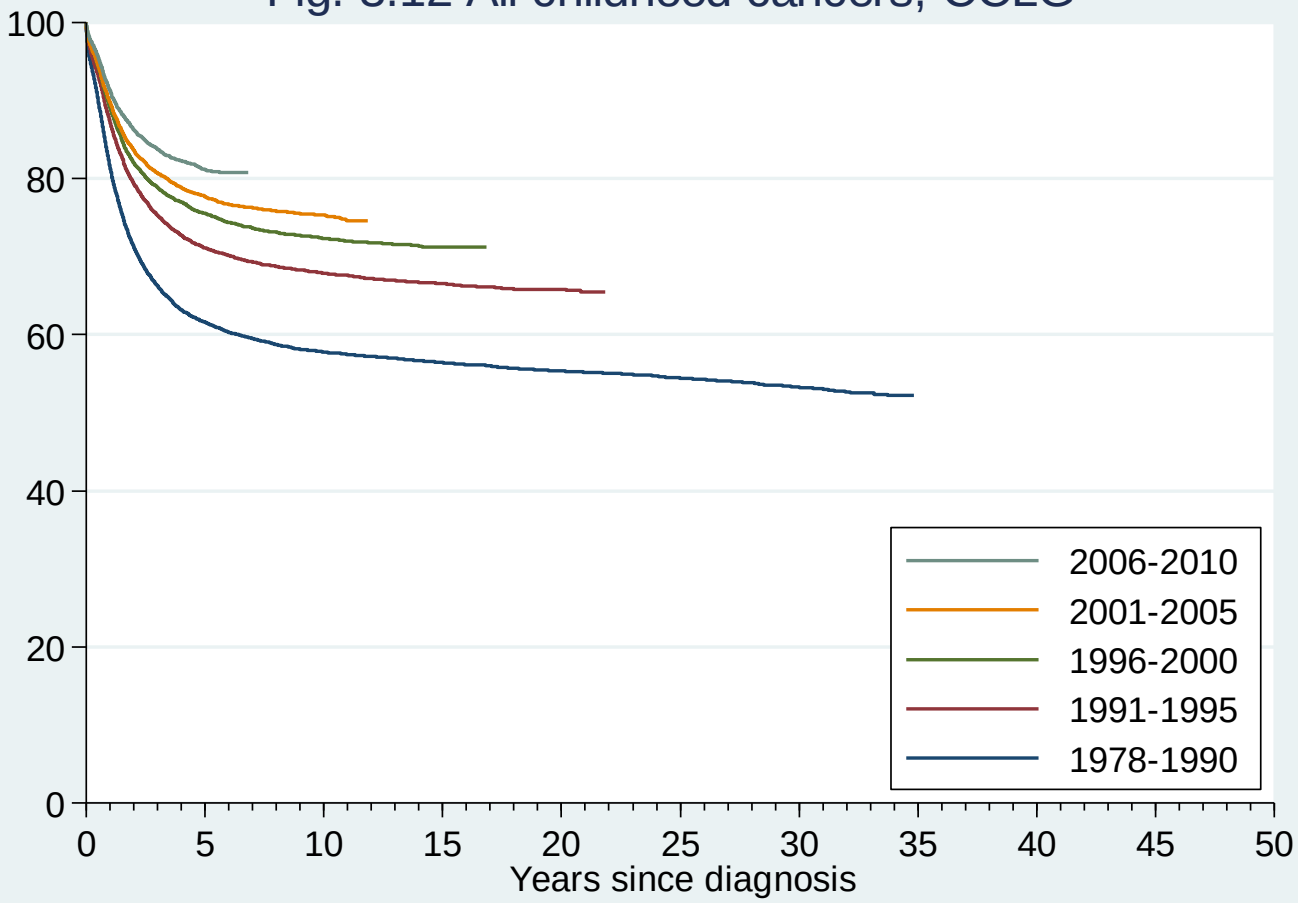


Fig. 3.13 Precursor-cell ALL, CCLG

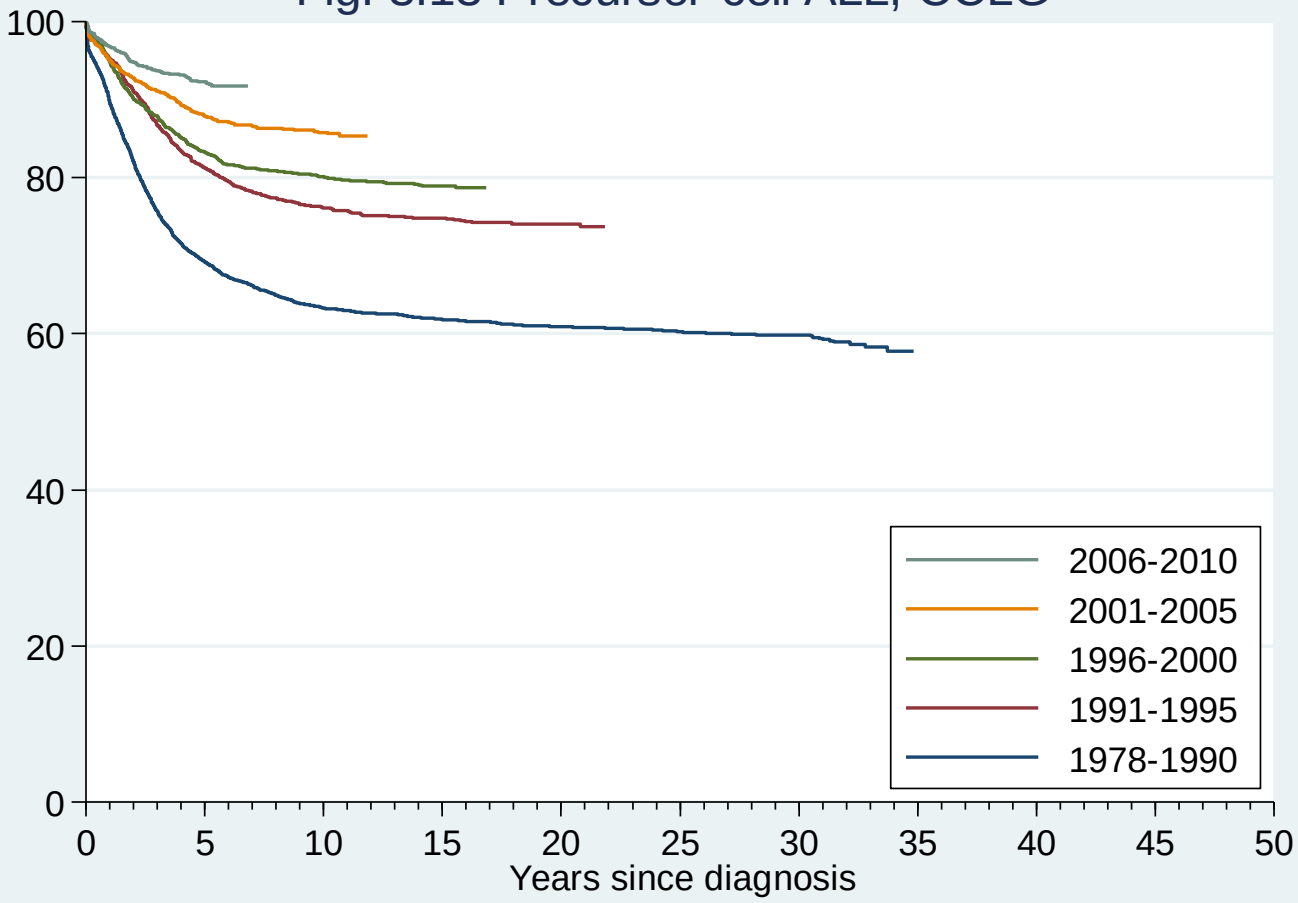


Fig. 3.14 Mature B-cell leukaemia, CCLG

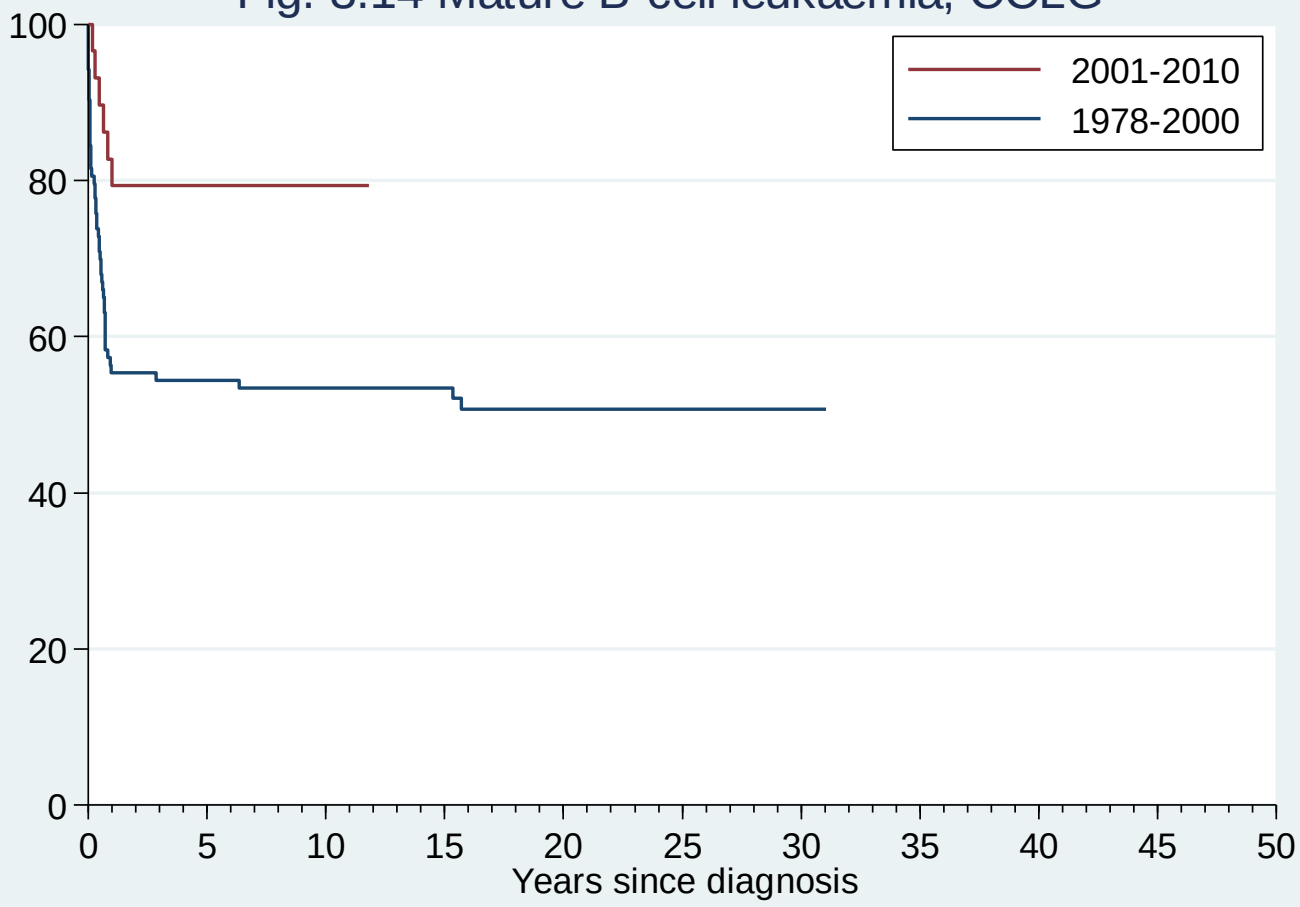


Fig. 3.15 Acute myeloid leukaemia, CCLG

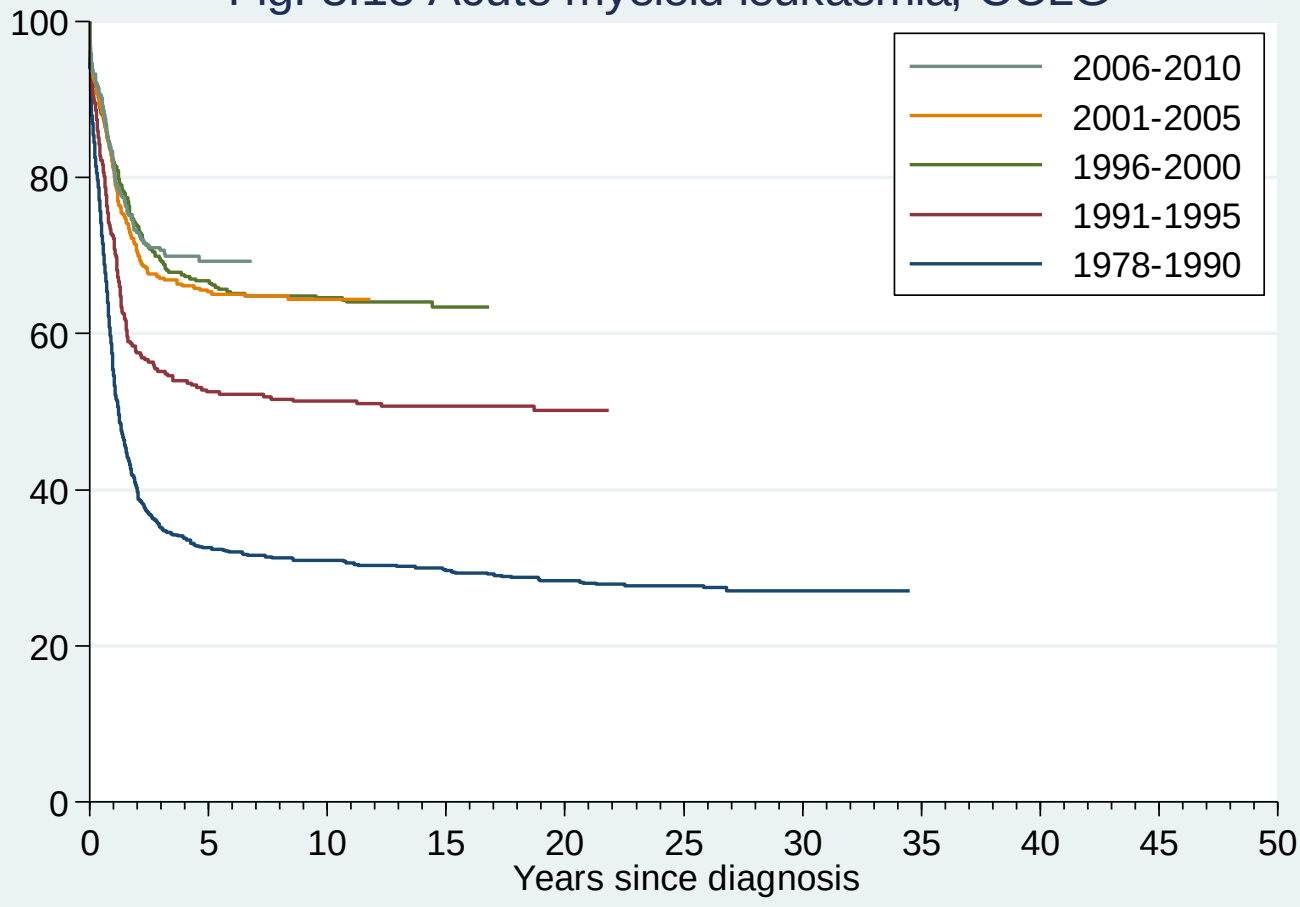


Fig. 3.16 Chronic myeloid leukaemia, CCLG

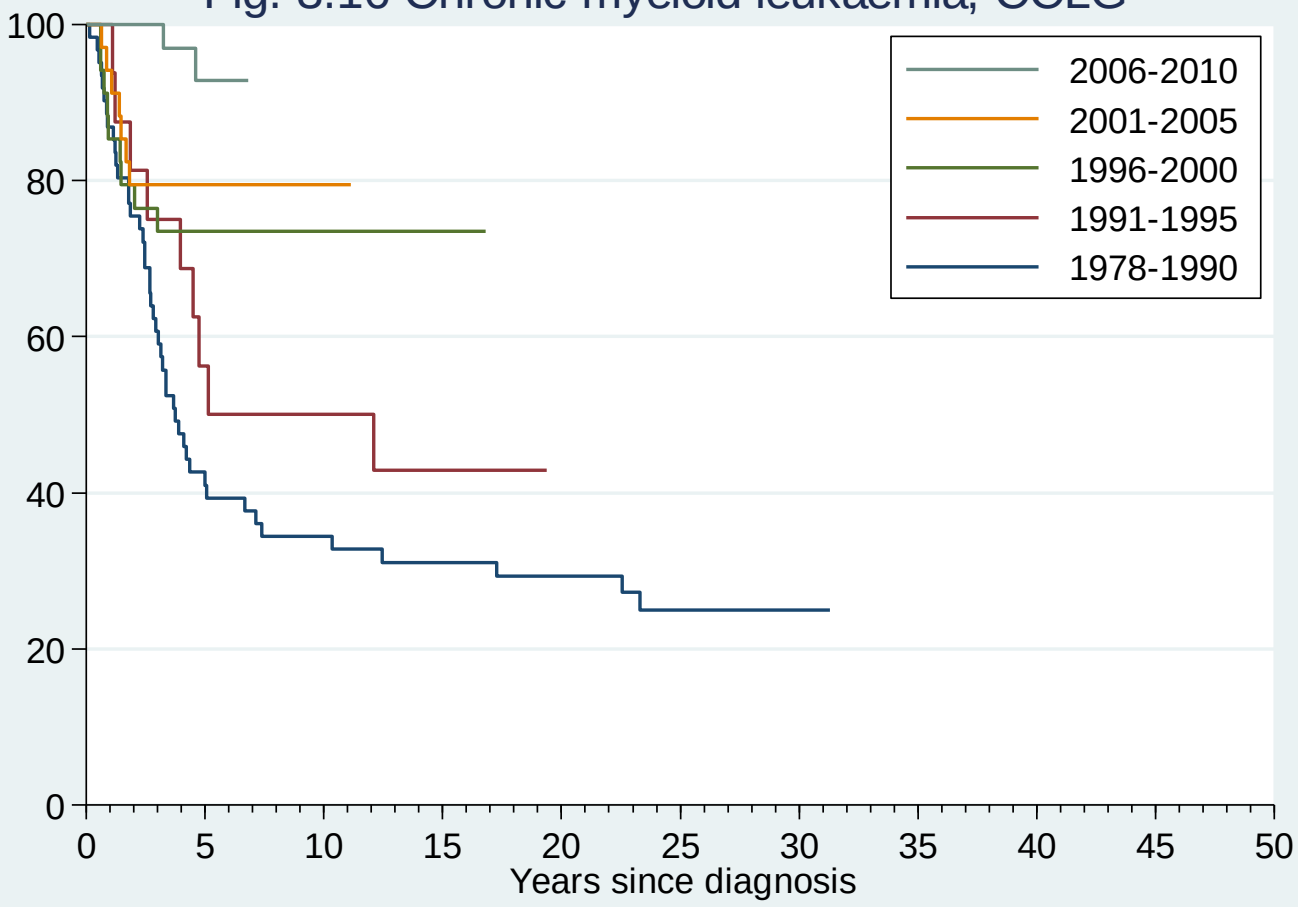


Fig. 3.17 Myelodysplasia, CCLG

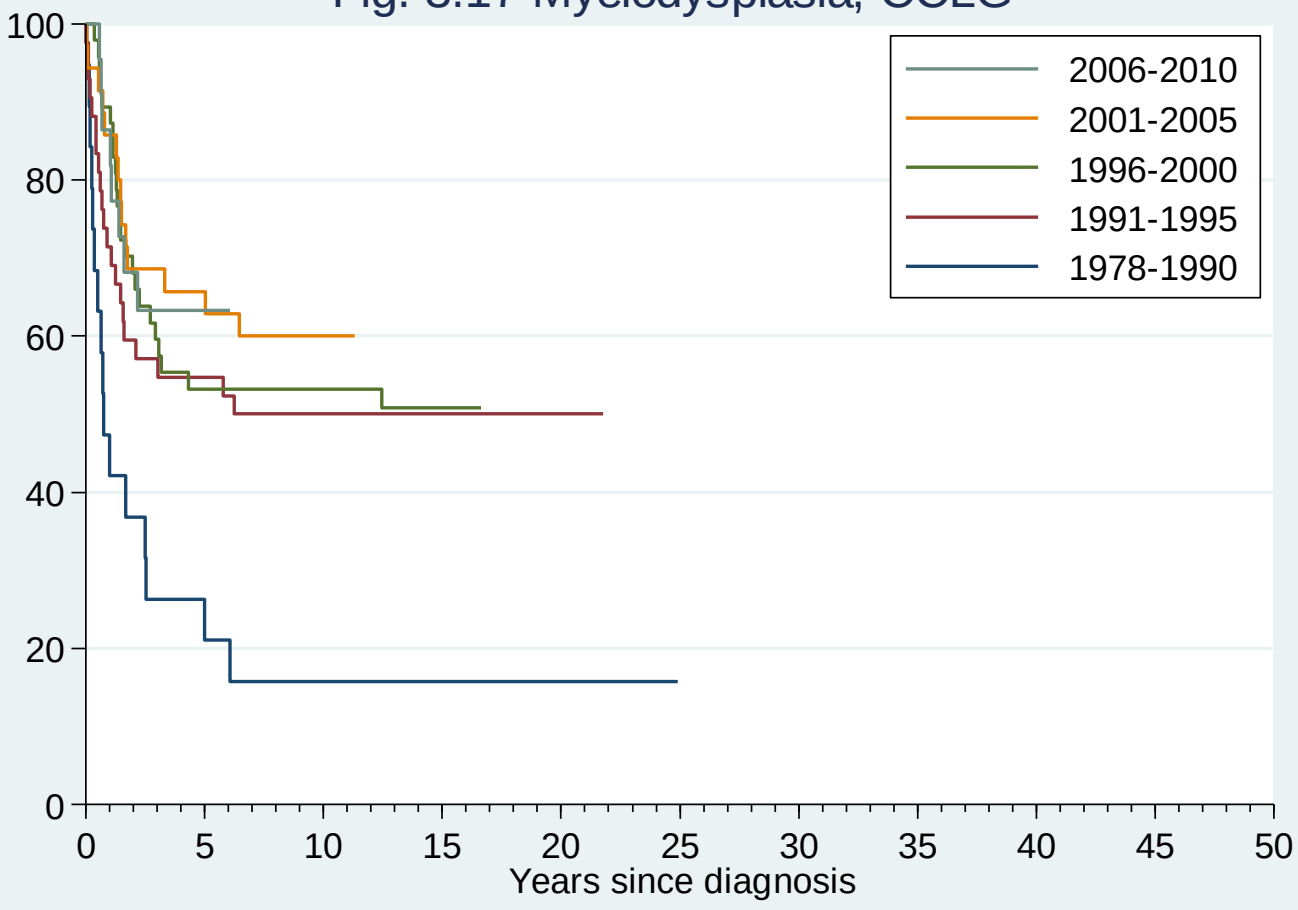


Fig. 3.18 JMML and CMML, CCLG

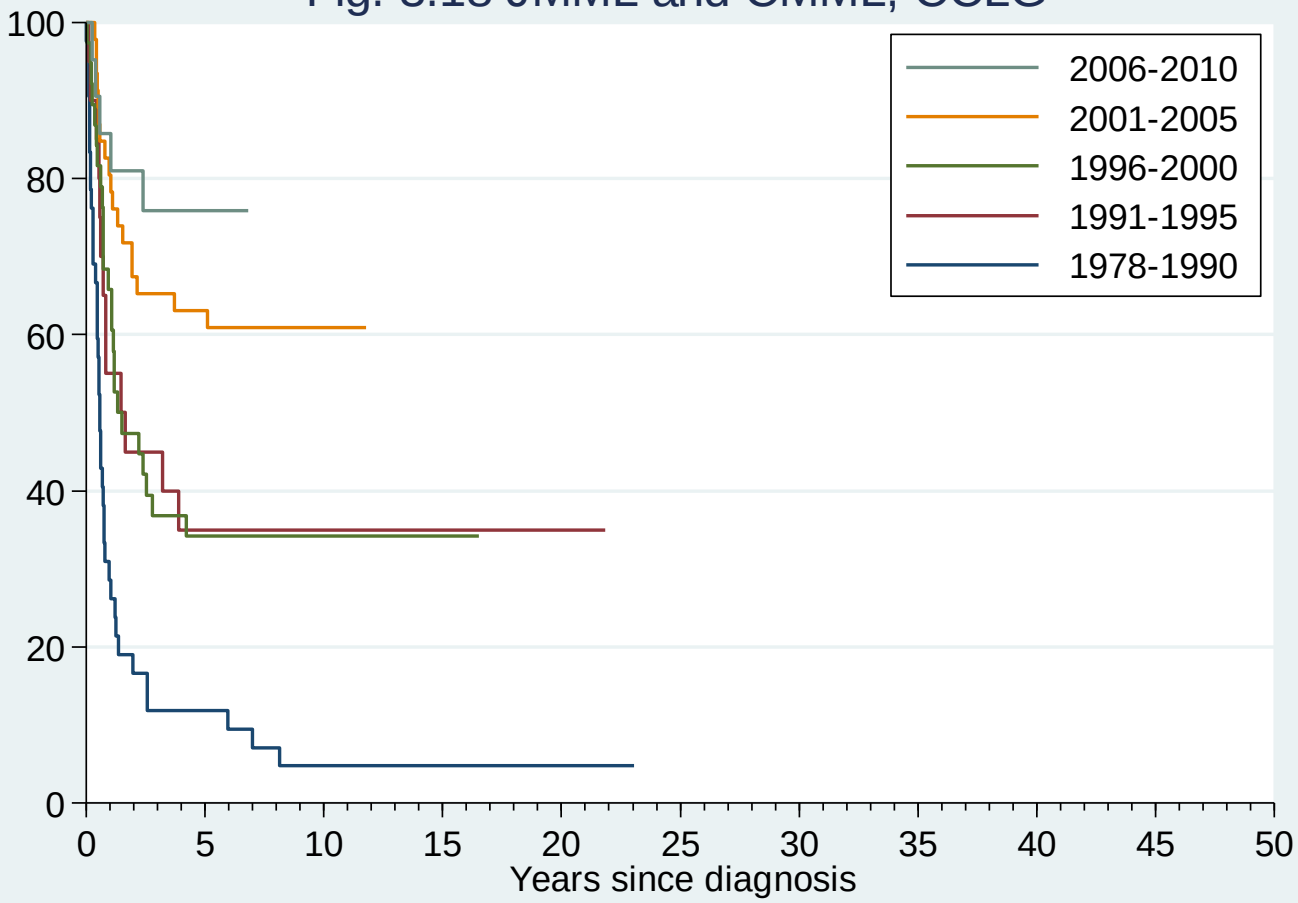


Fig. 3.19 Other and unspecified leukaemia, CCLG

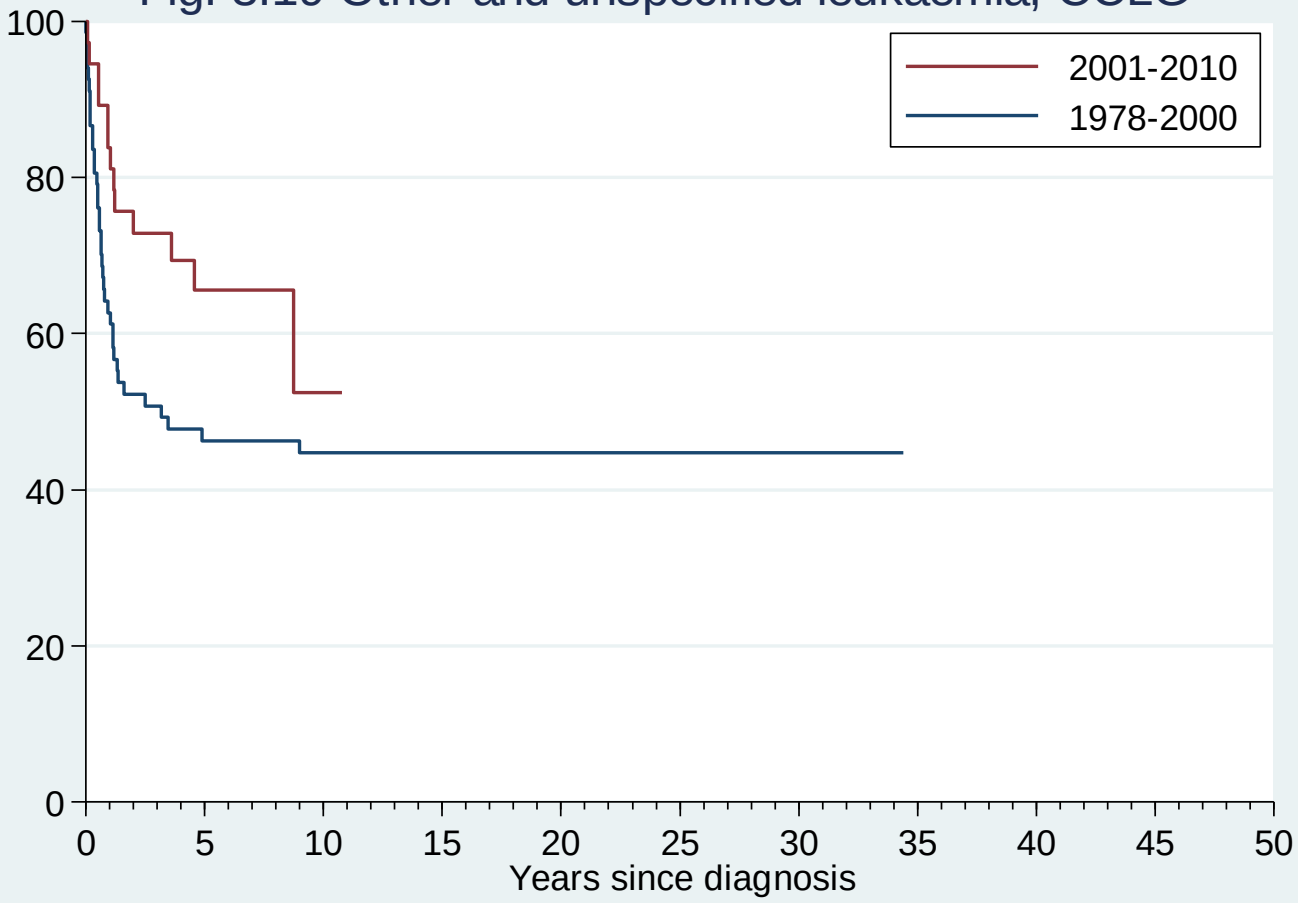


Fig. 3.20 Hodgkin lymphoma, CCLG

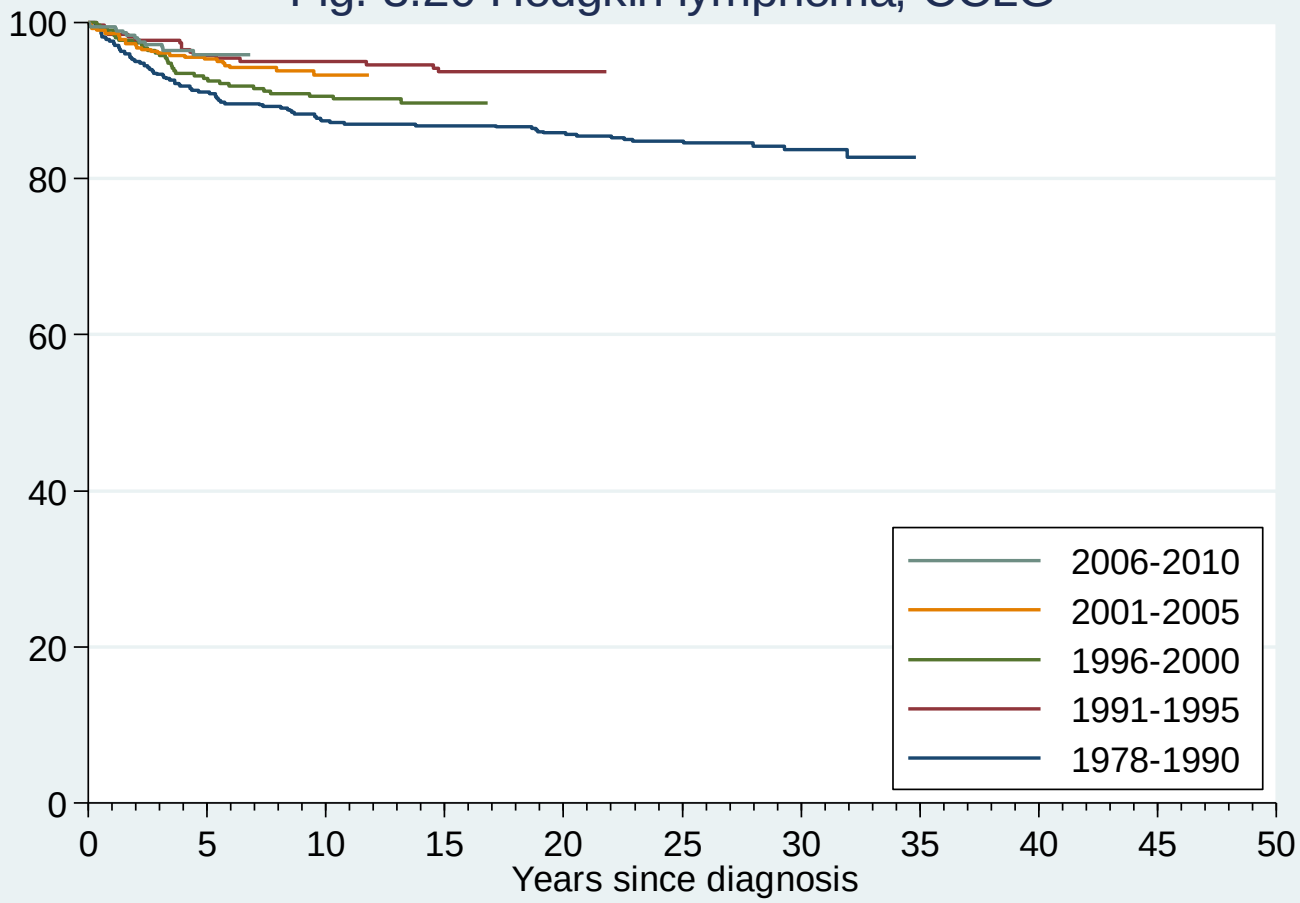


Fig. 3.21 Non-Hodgkin lymphoma, CCLG

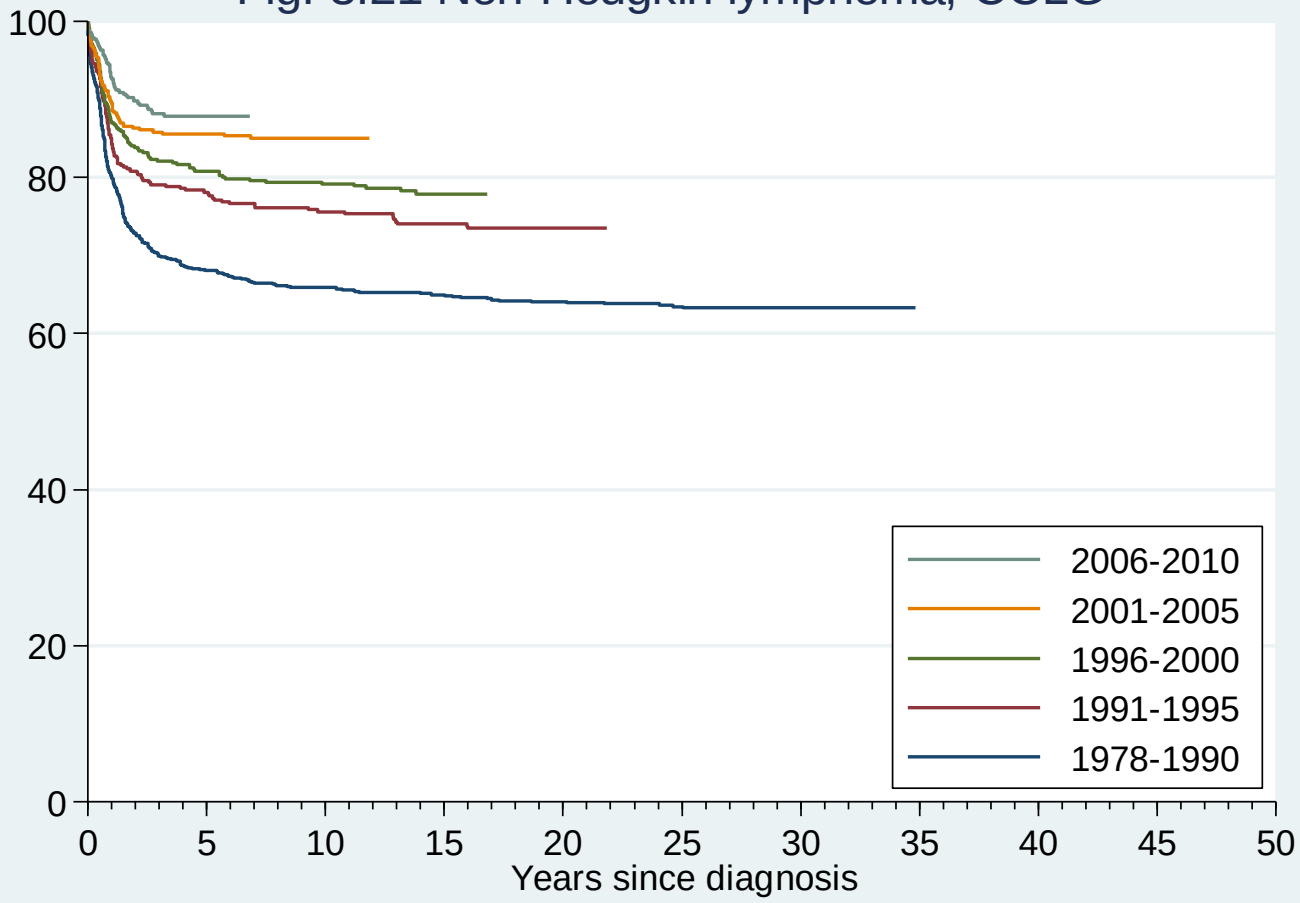


Fig. 3.22 Ependymoma, CCLG

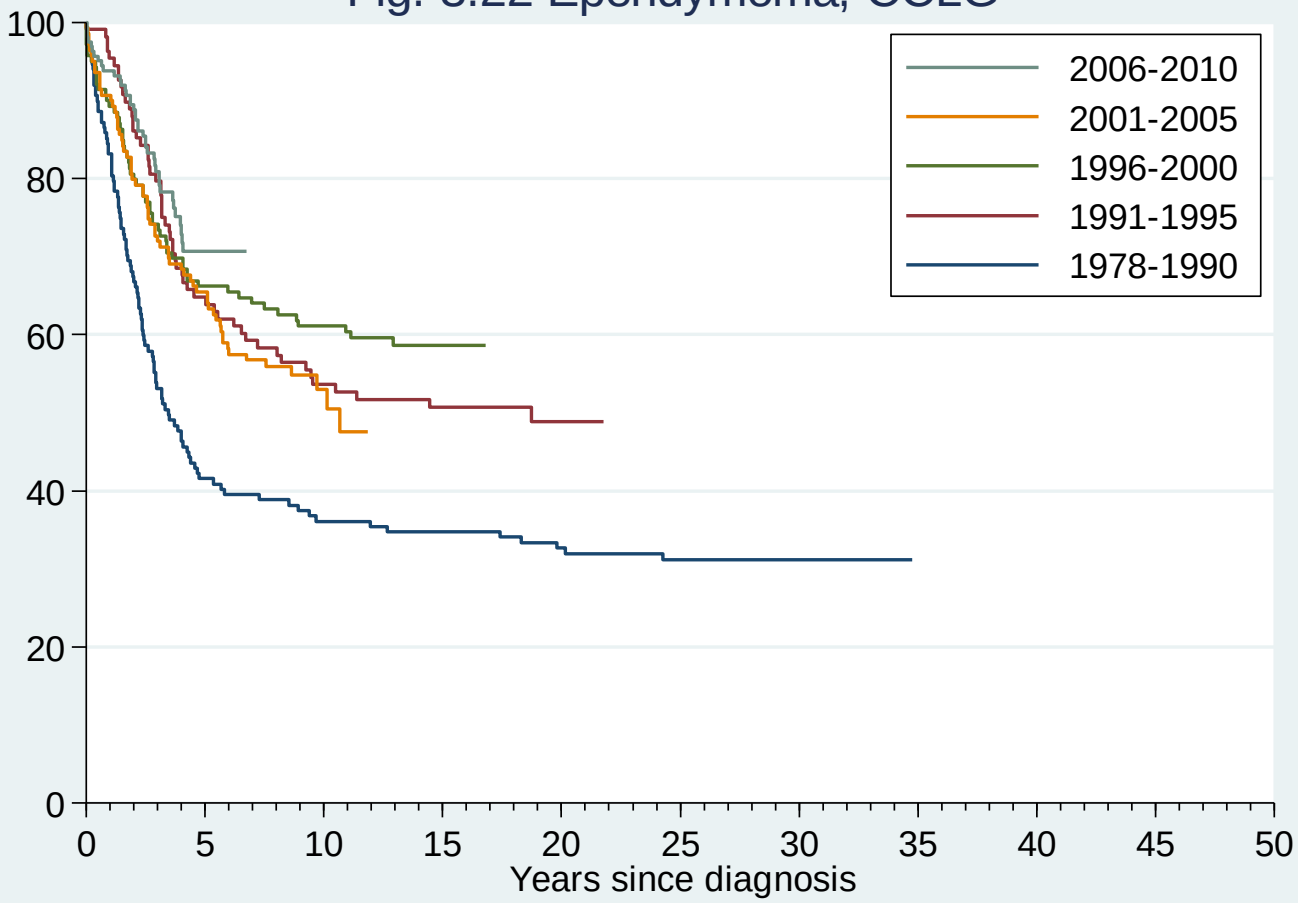


Fig. 3.23 Choroid plexus papilloma, CCLG

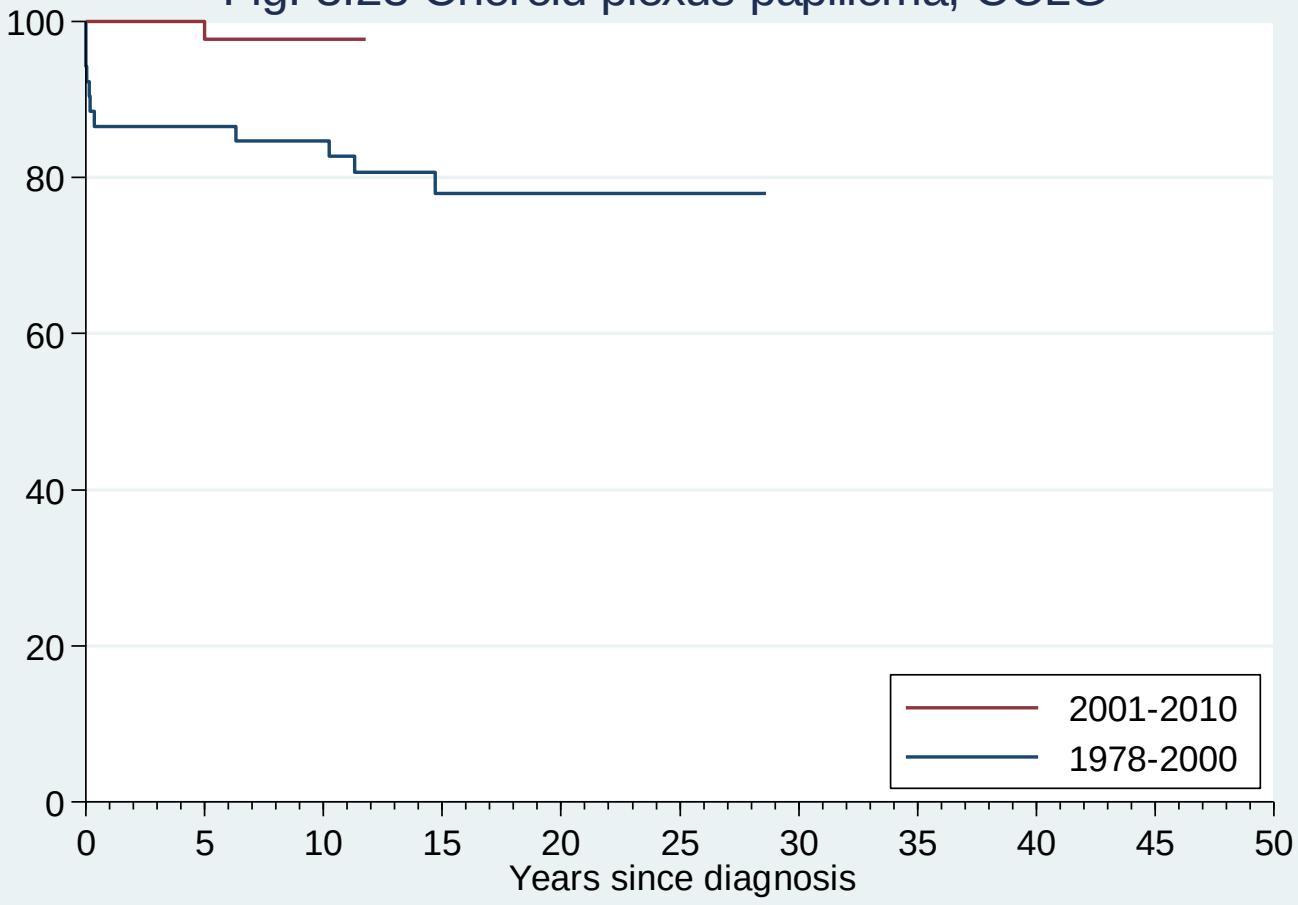


Fig. 3.24 Choroid plexus carcinoma, CCLG

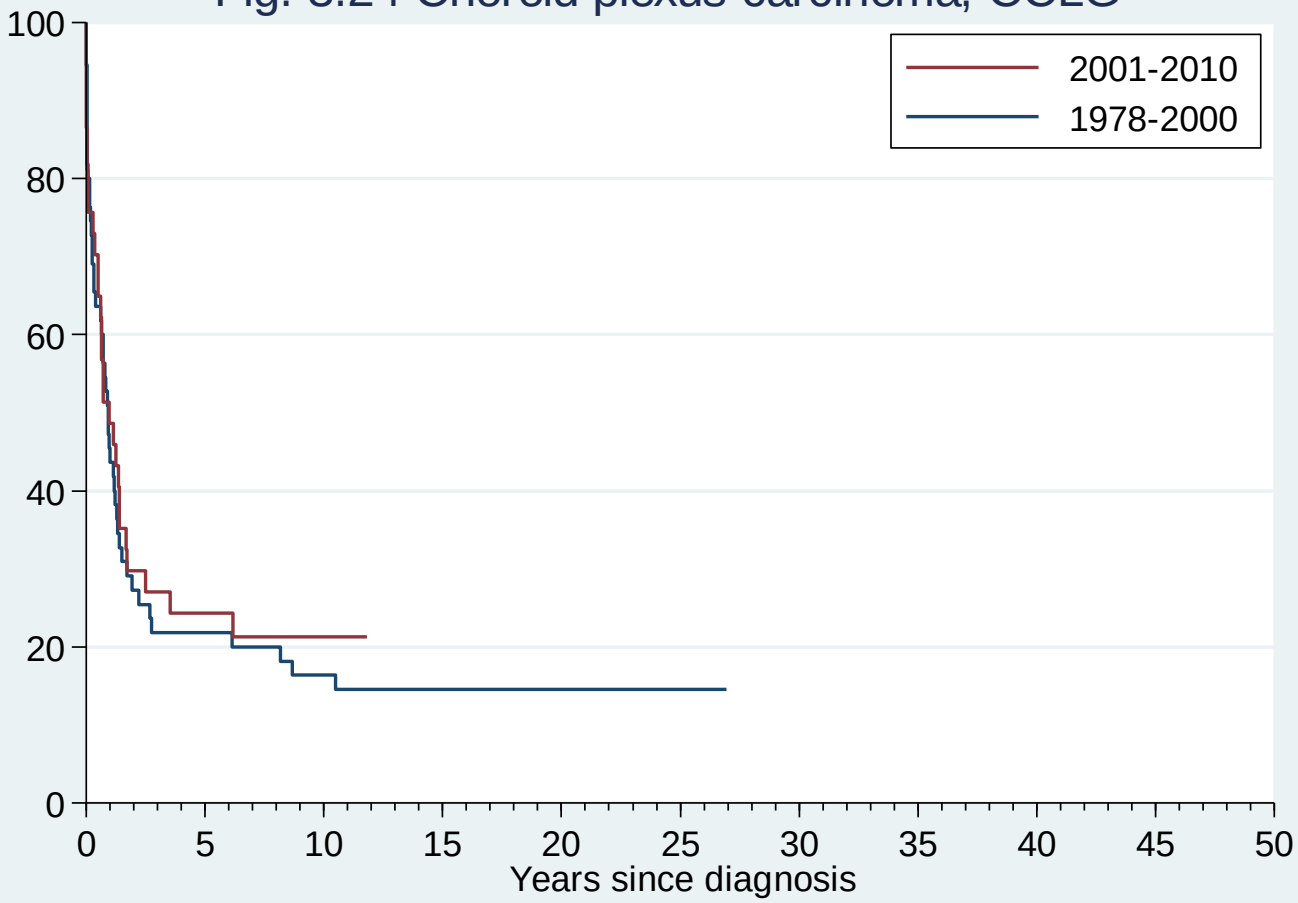


Fig. 3.25 Low-grade astrocytoma, CCLG

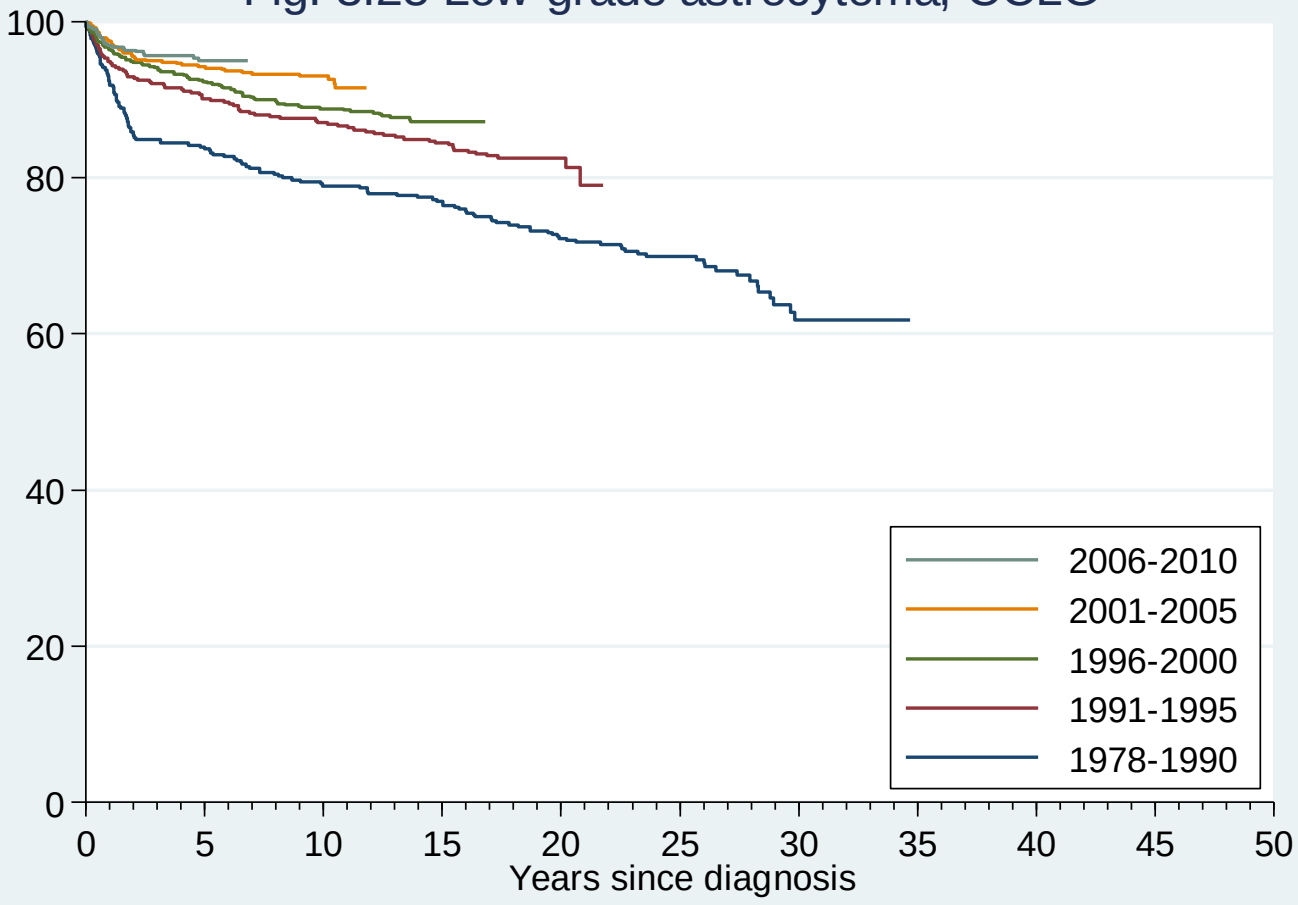


Fig. 3.26 High-grade astrocytoma, CCLG

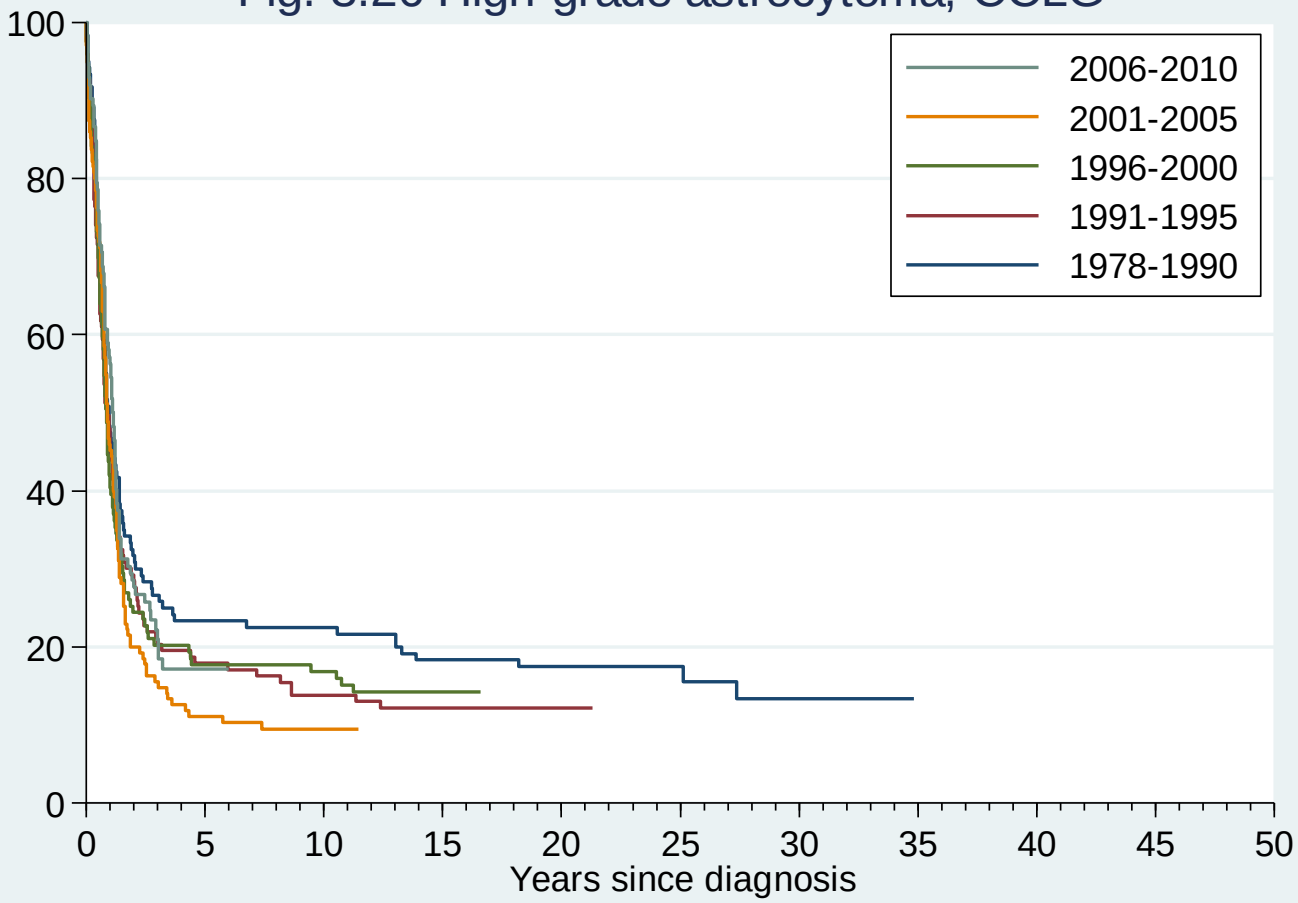


Fig. 3.27 Unspecified astrocytoma, CCLG

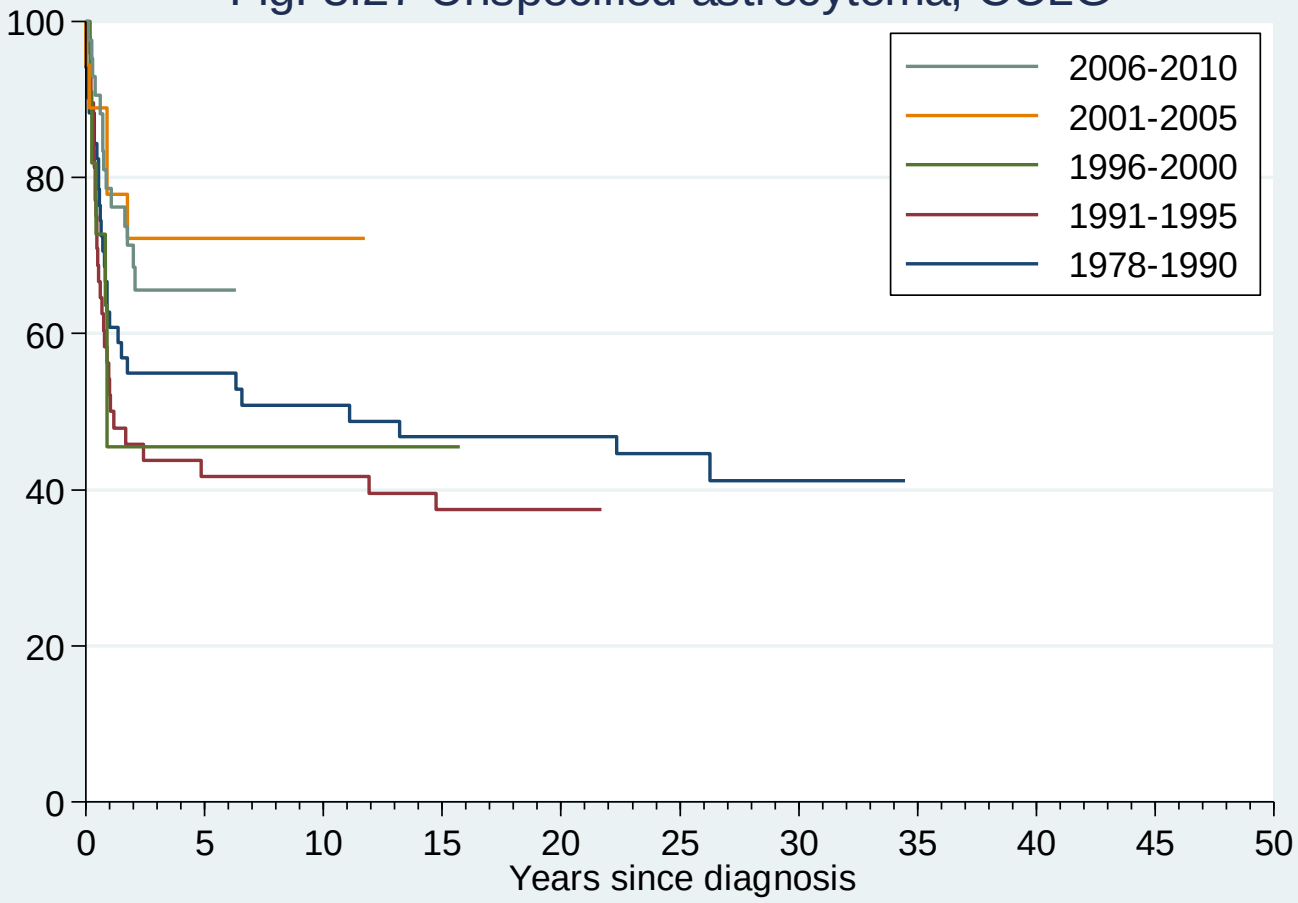


Fig. 3.28 Medulloblastoma, CCLG

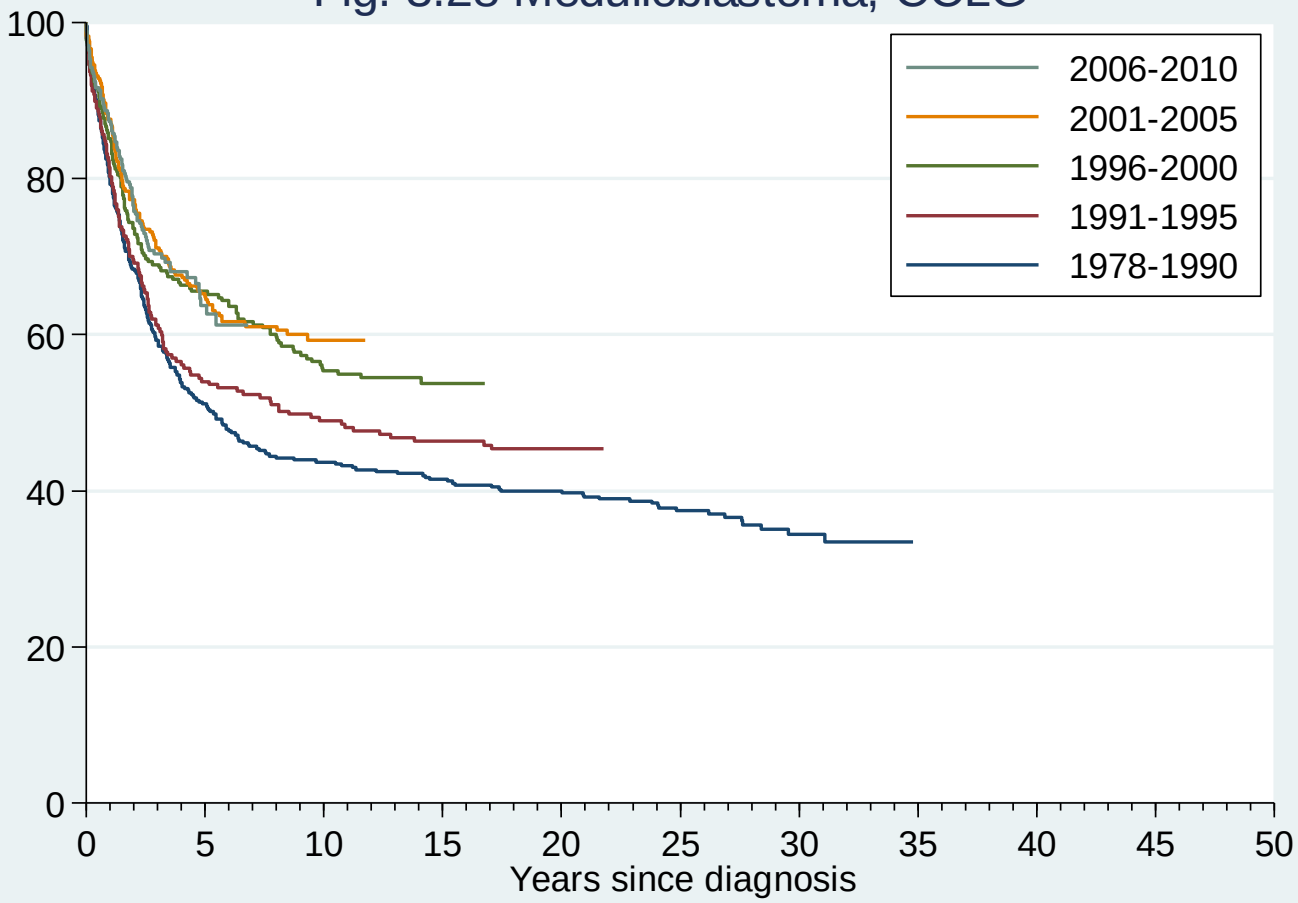


Fig. 3.29 CNS PNET, CCLG

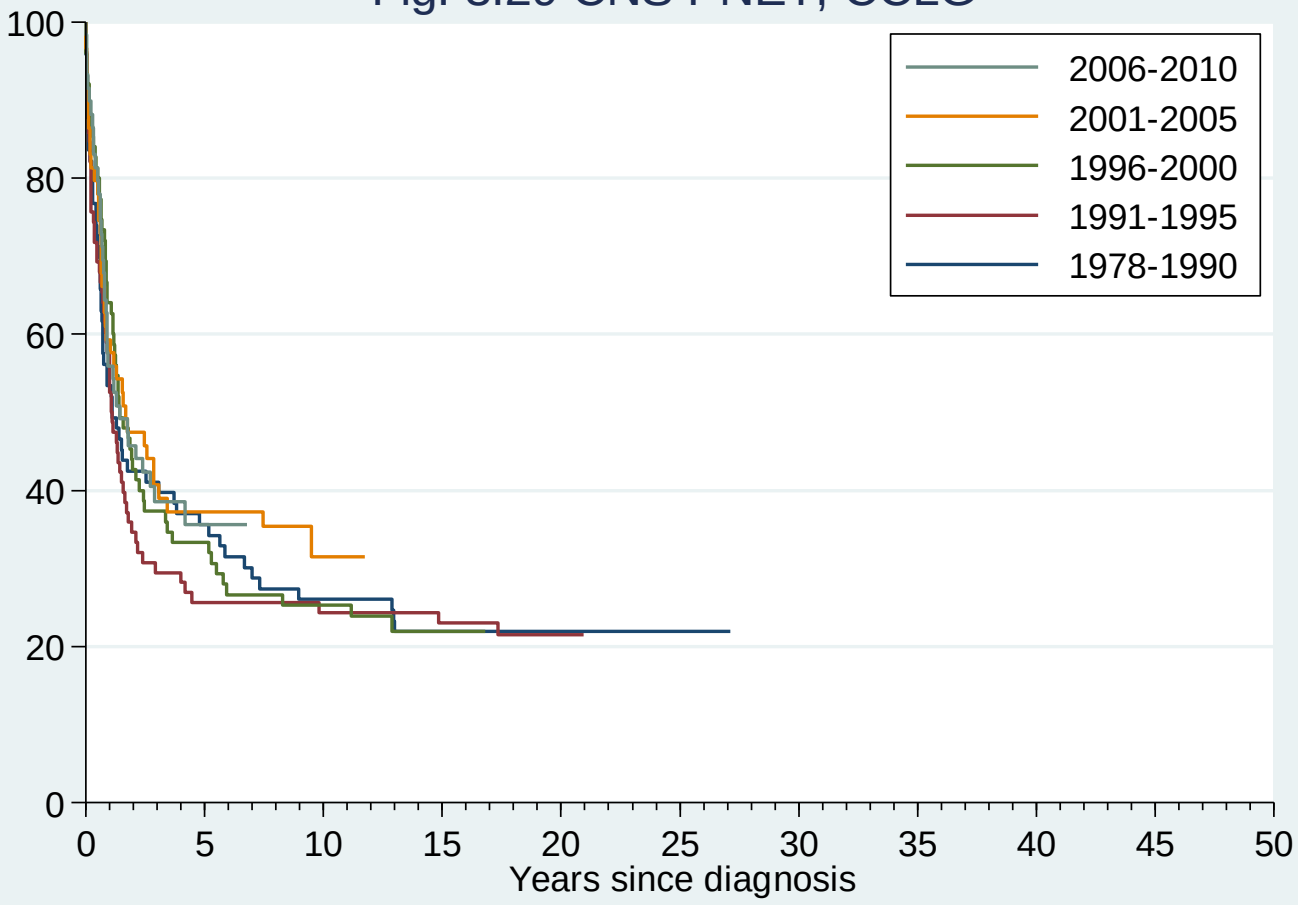


Fig. 3.30 Atypical teratoid/rhabdoid tumour, CCLG

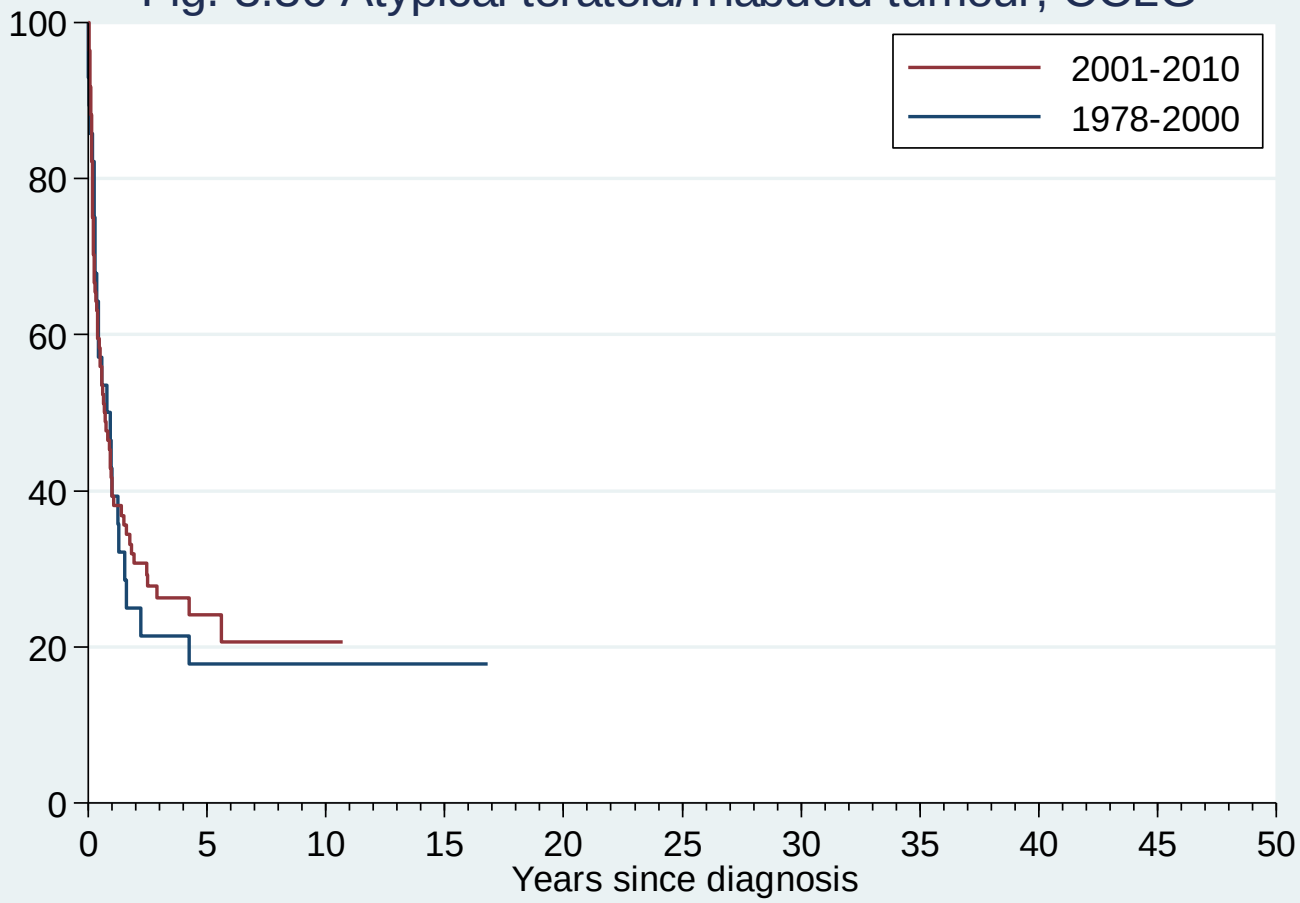


Fig. 3.31 Oligodendroglioma, CCLG

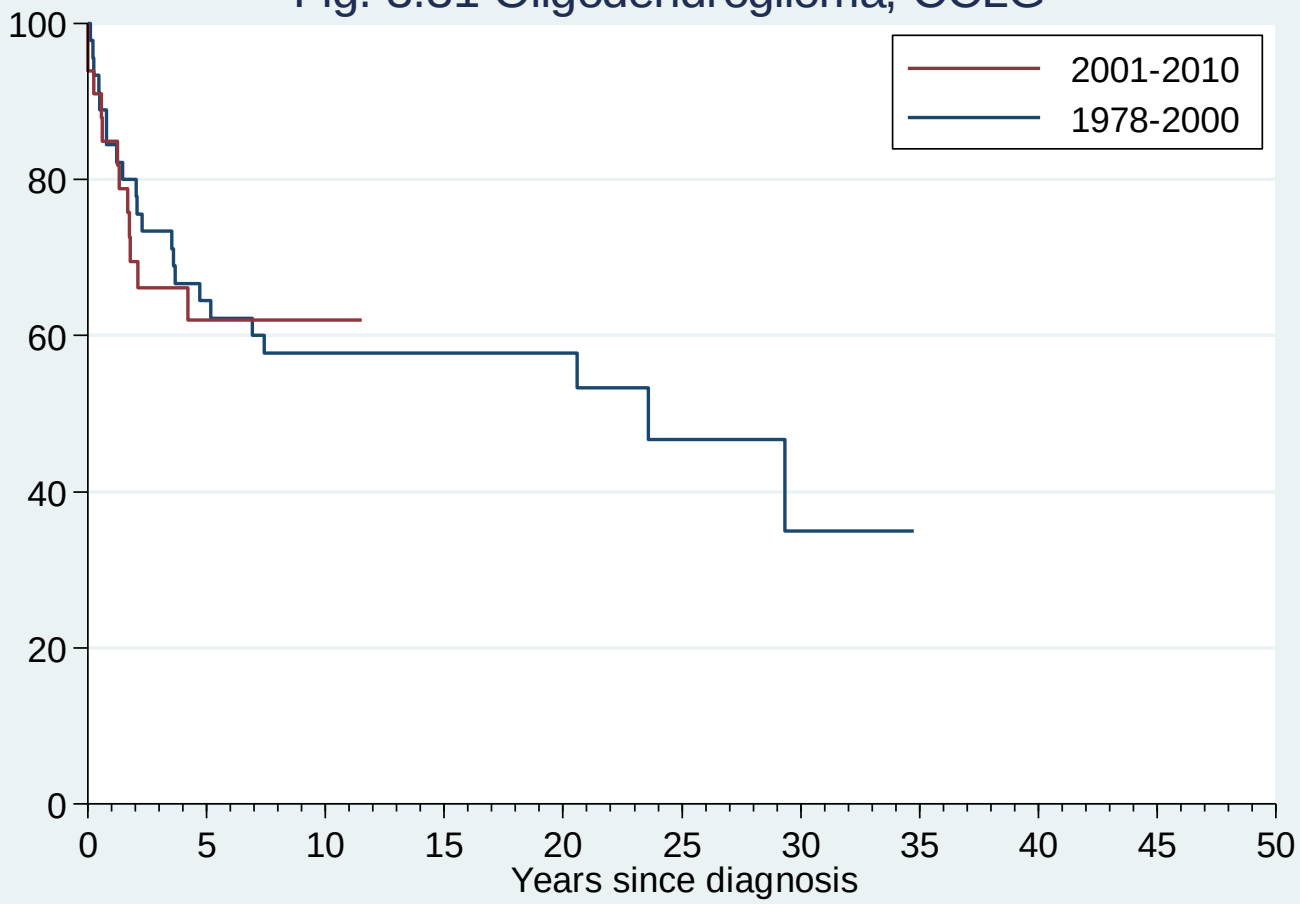


Fig. 3.32 Other glioma, CCLG

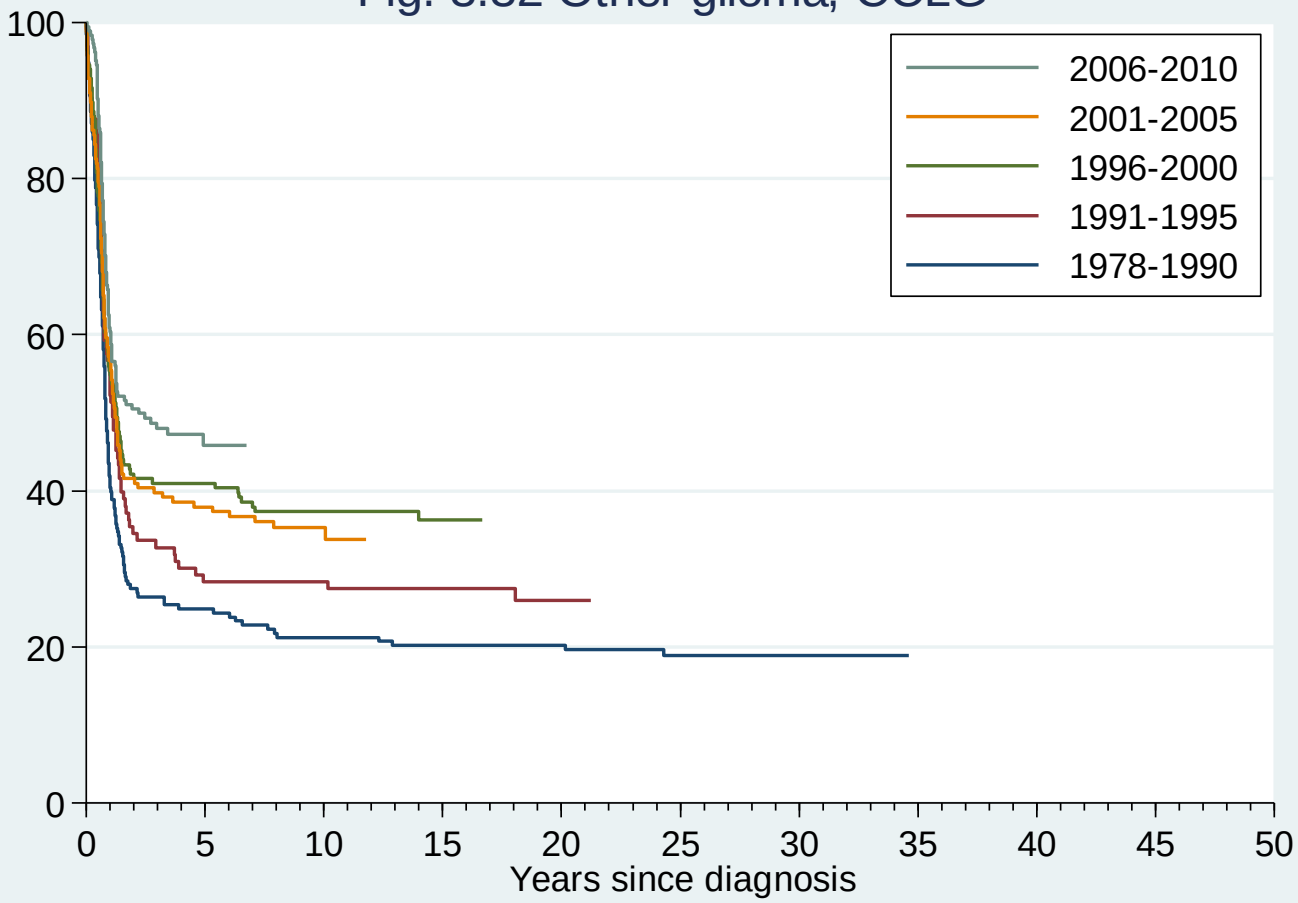


Fig. 3.33 Pituitary carcinoma and adenoma, CCLG

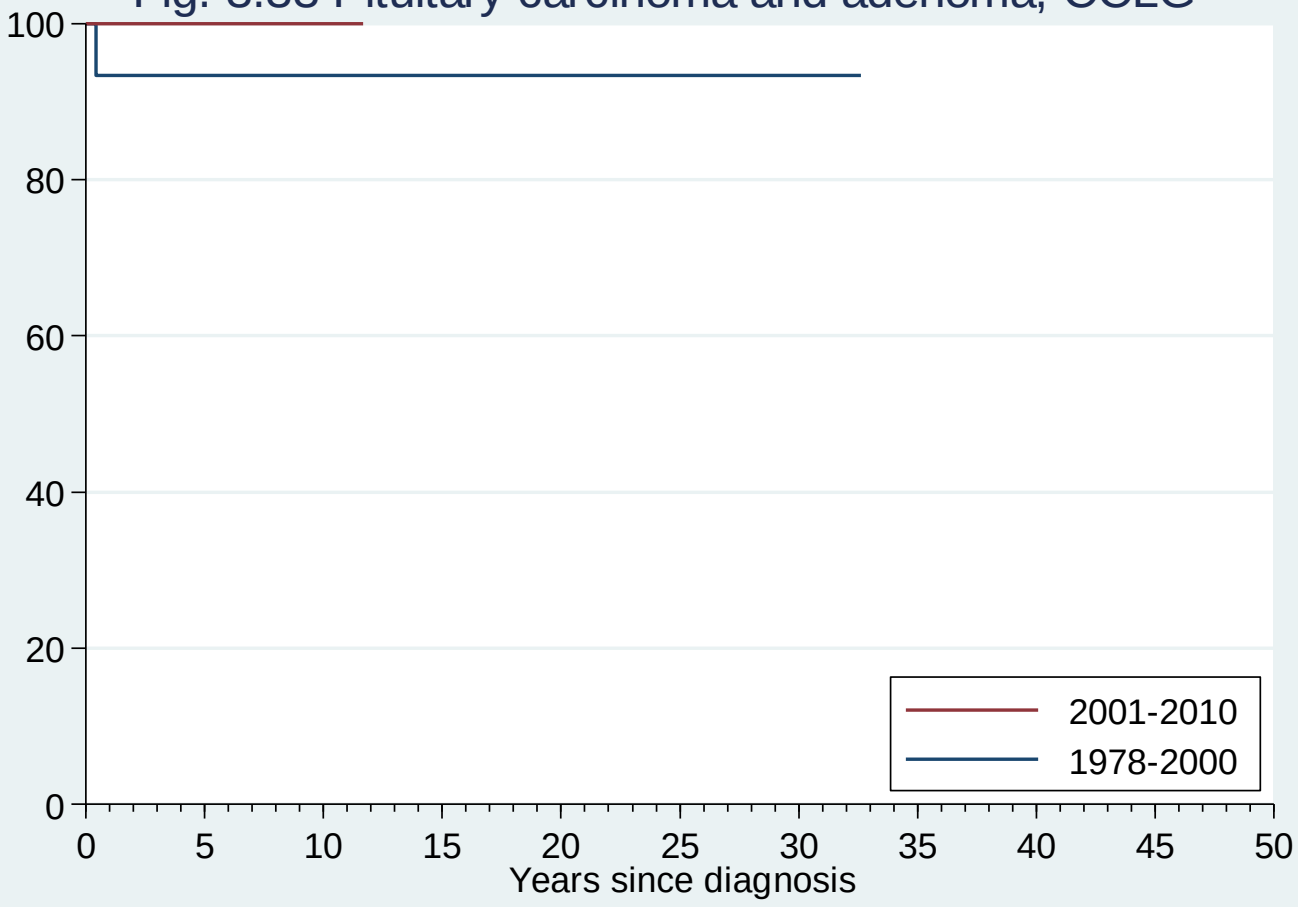


Fig. 3.34 Craniopharyngioma, CCLG

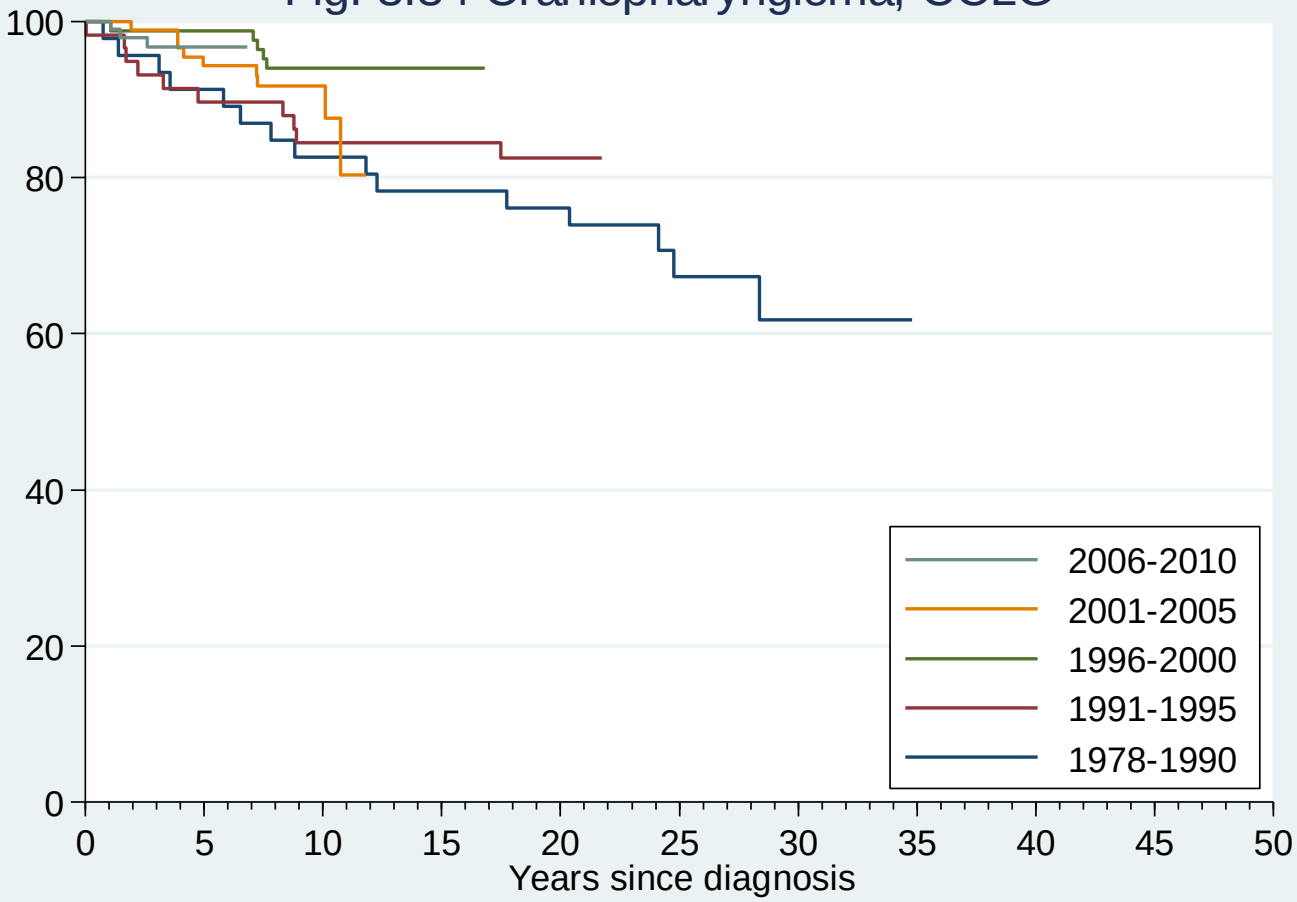


Fig. 3.35 Pinealoma and pineocytoma, CCLG

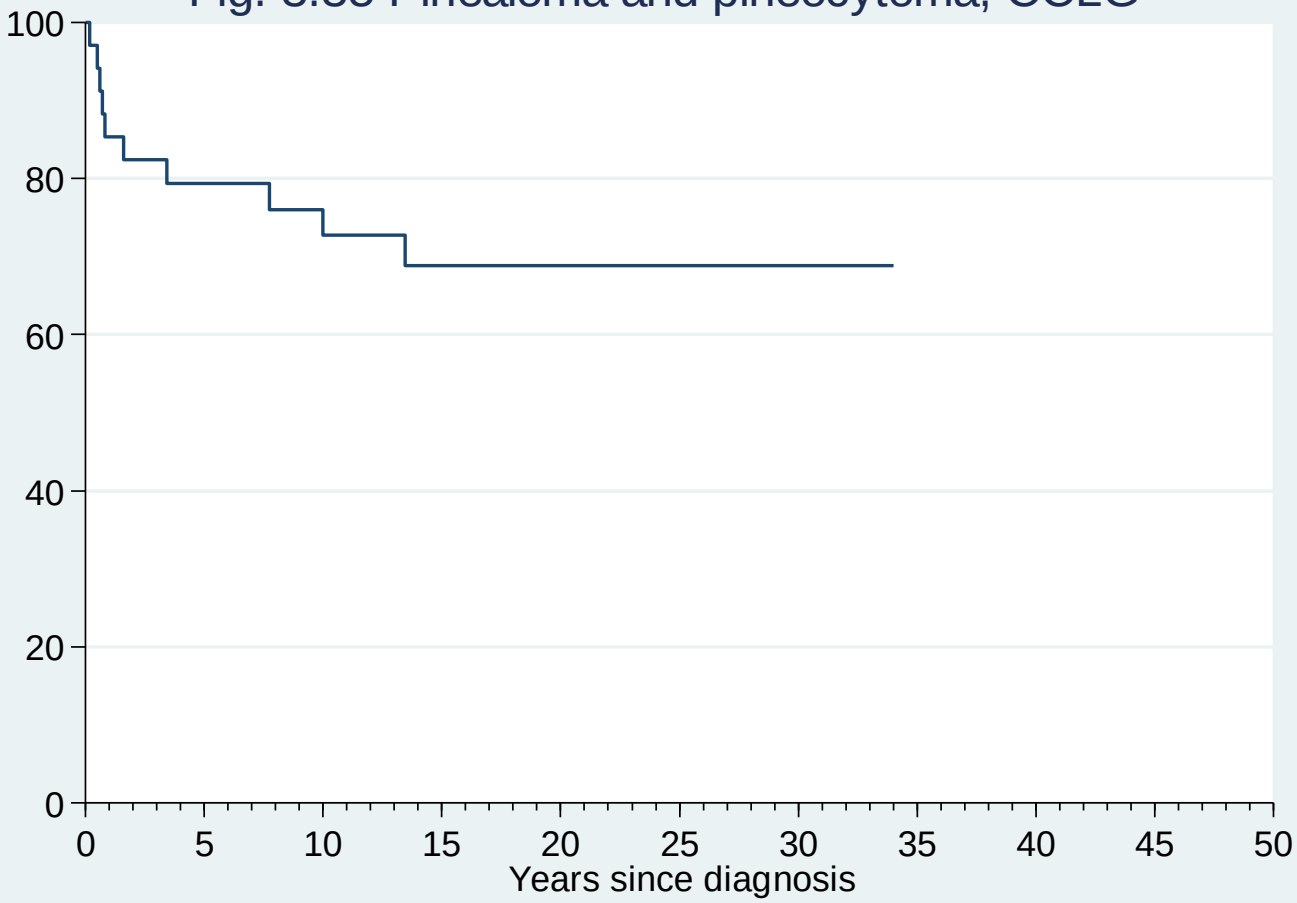


Fig. 3.36 Pineoblastoma, CCLG

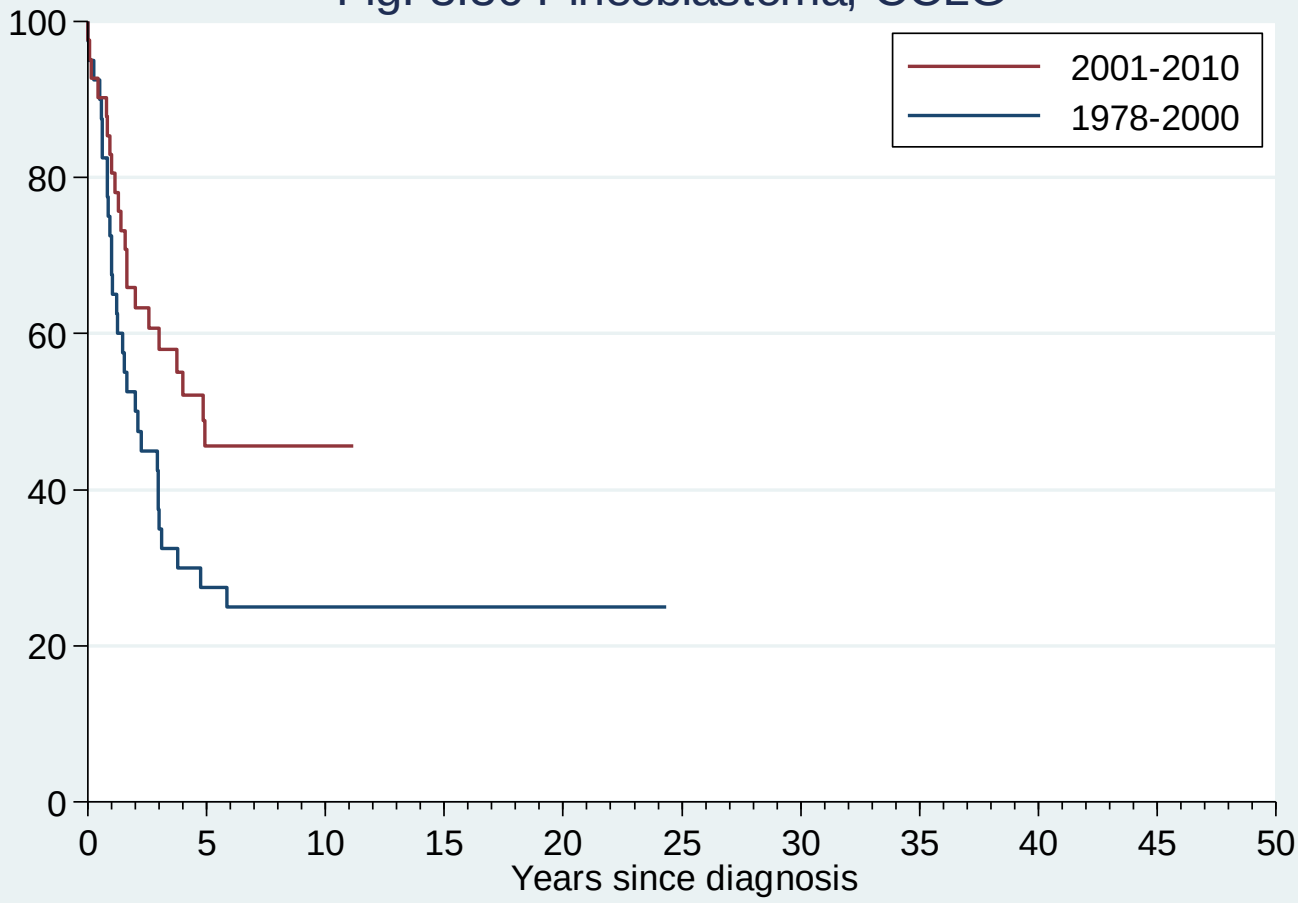


Fig. 3.37 Ganglioglioma, CCLG

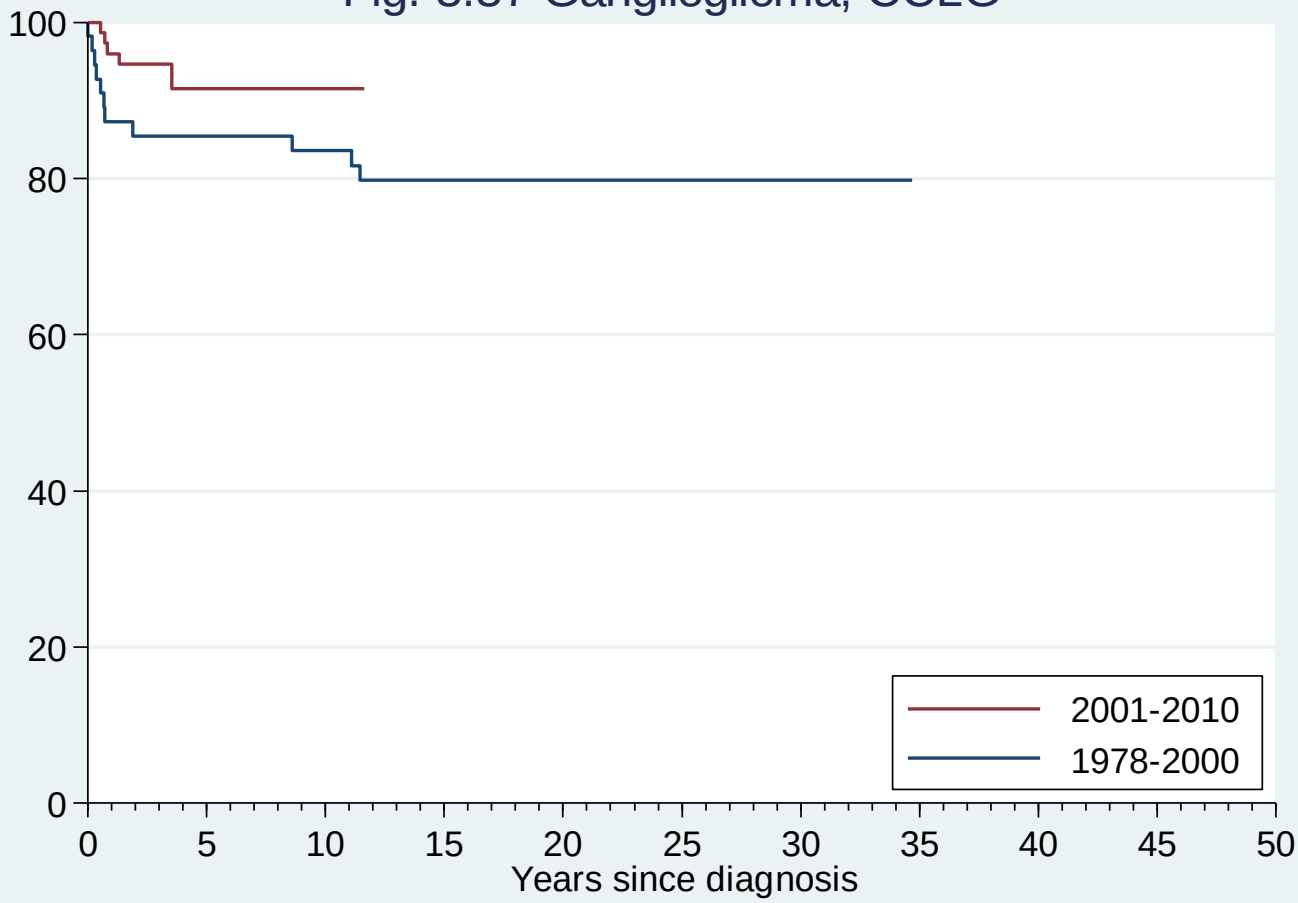


Fig. 3.38 Desmoplastic infantile astrocytoma, CCLG

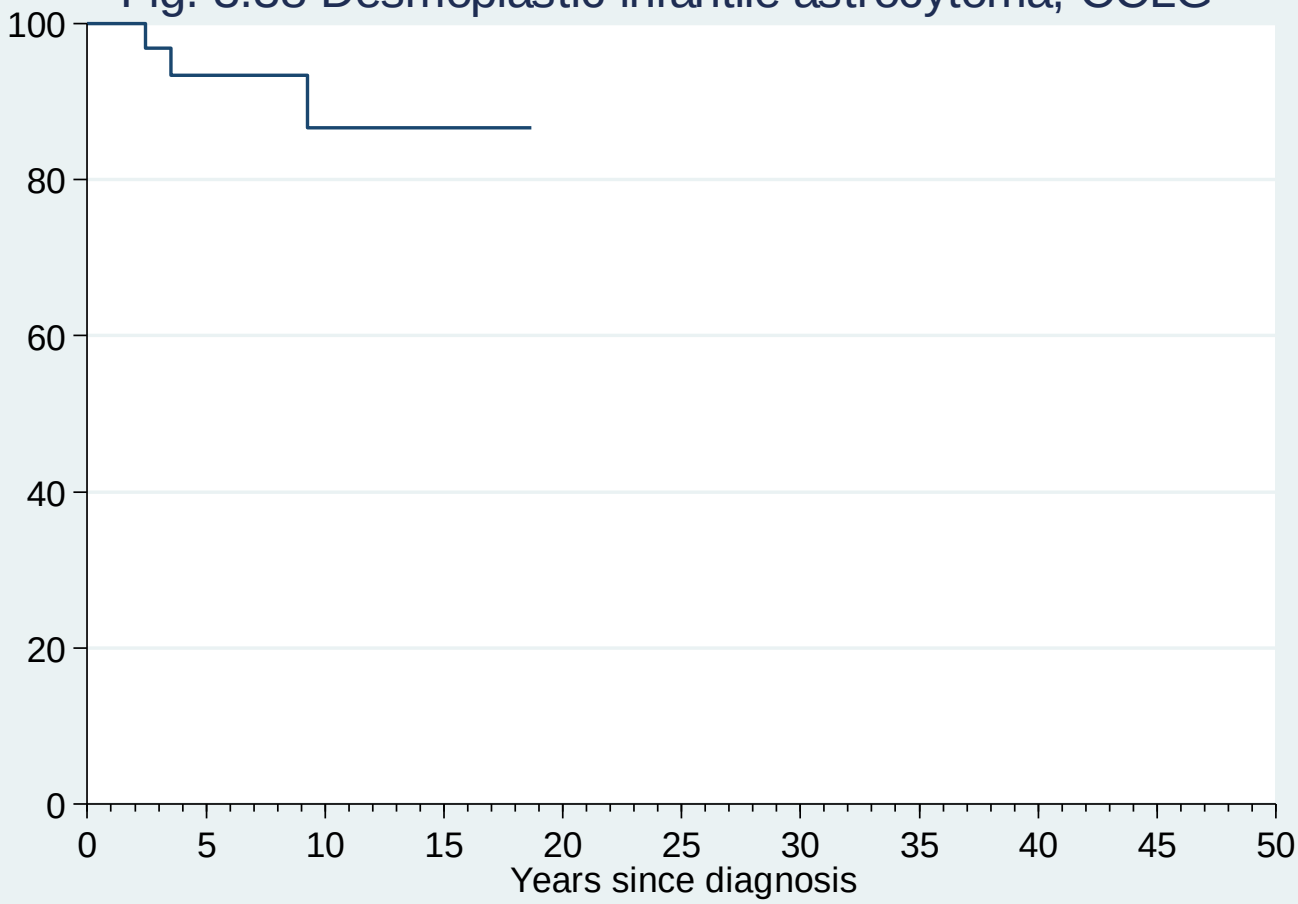


Fig. 3.39 Dysembryoplastic neuroepithelial tumour, CCLG

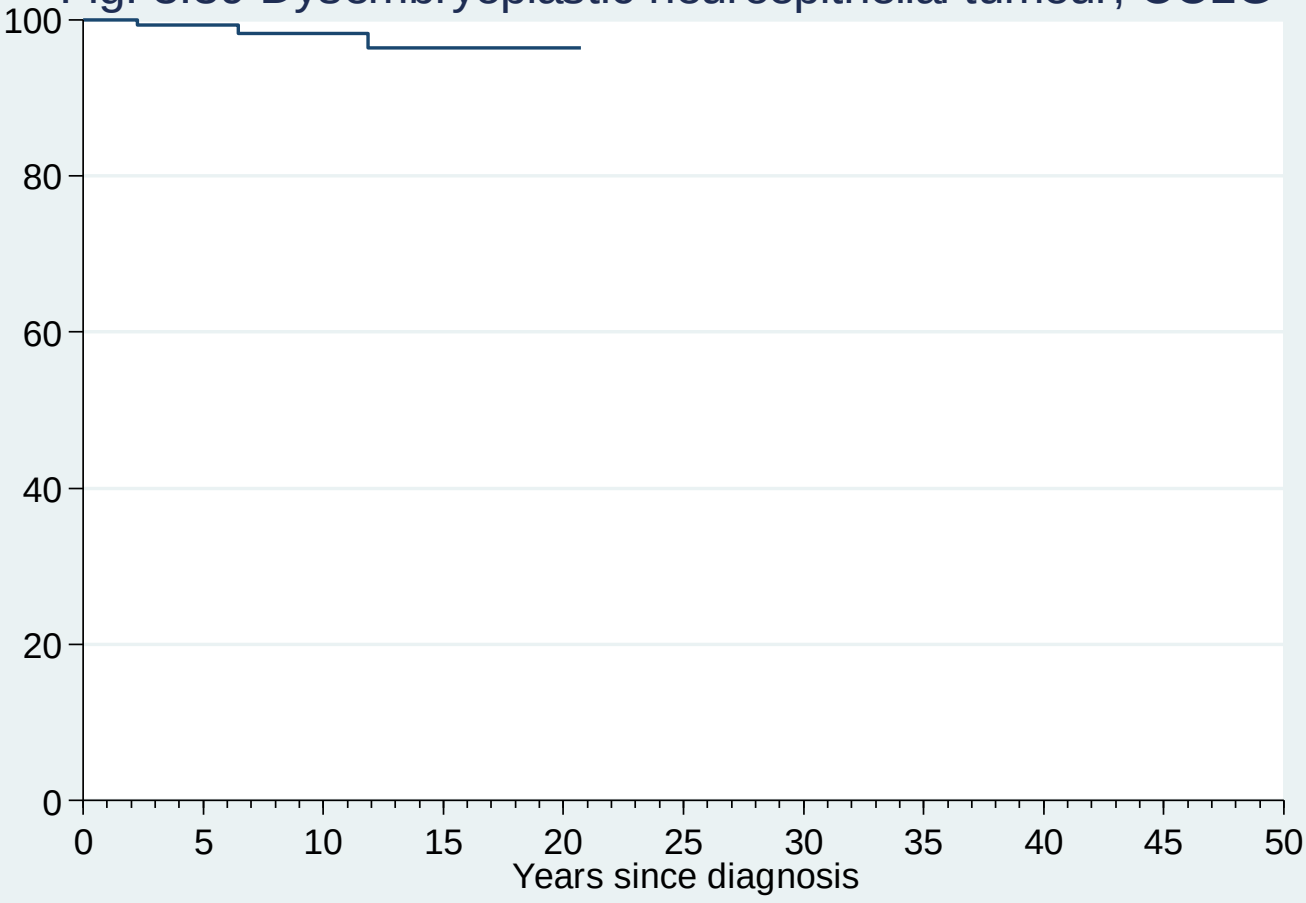


Fig. 3.40 Meningioma, CCLG

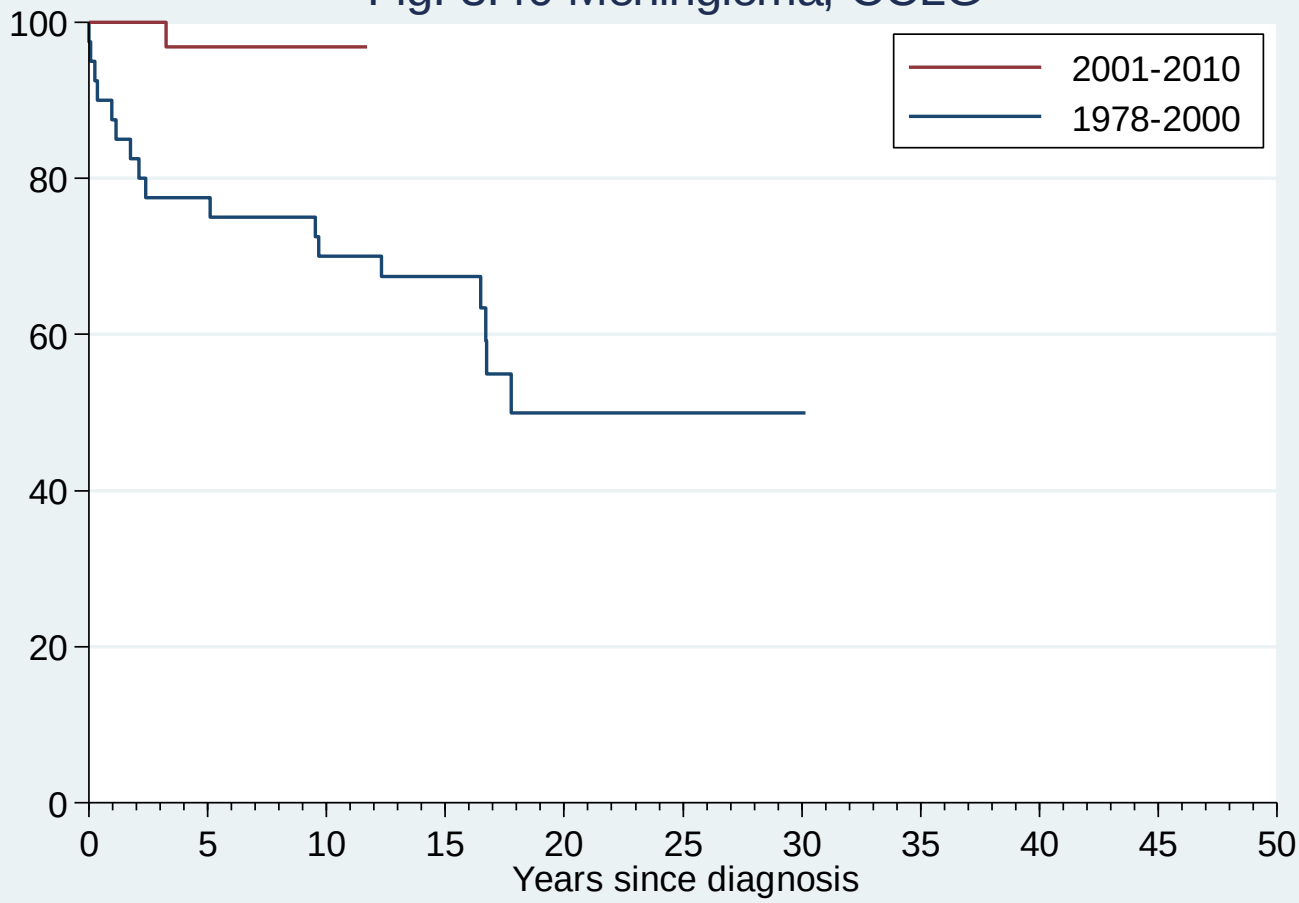


Fig. 3.41 Unspecified CNS tumours, CCLG

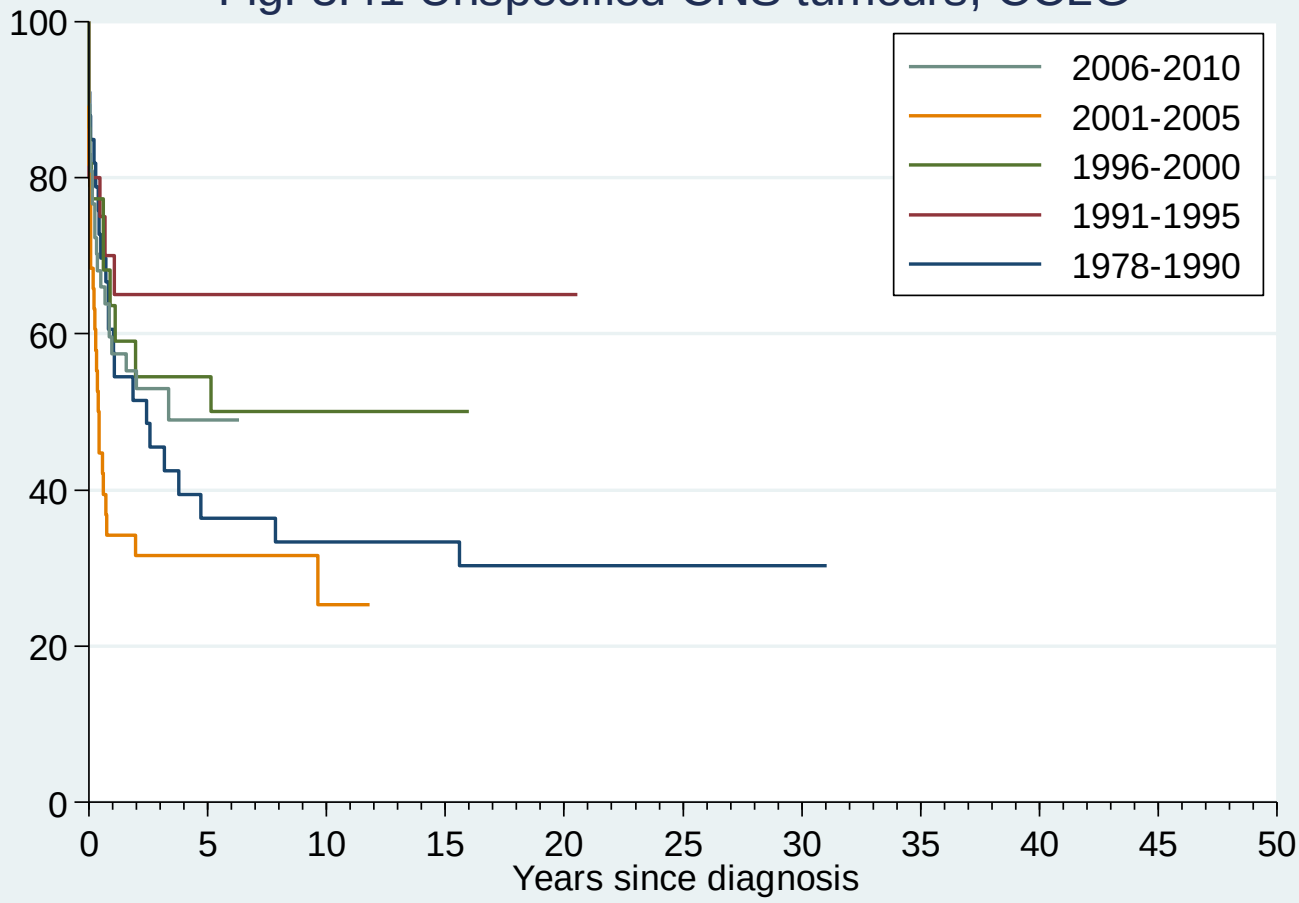


Fig. 3.42 Brain stem tumours, CCLG

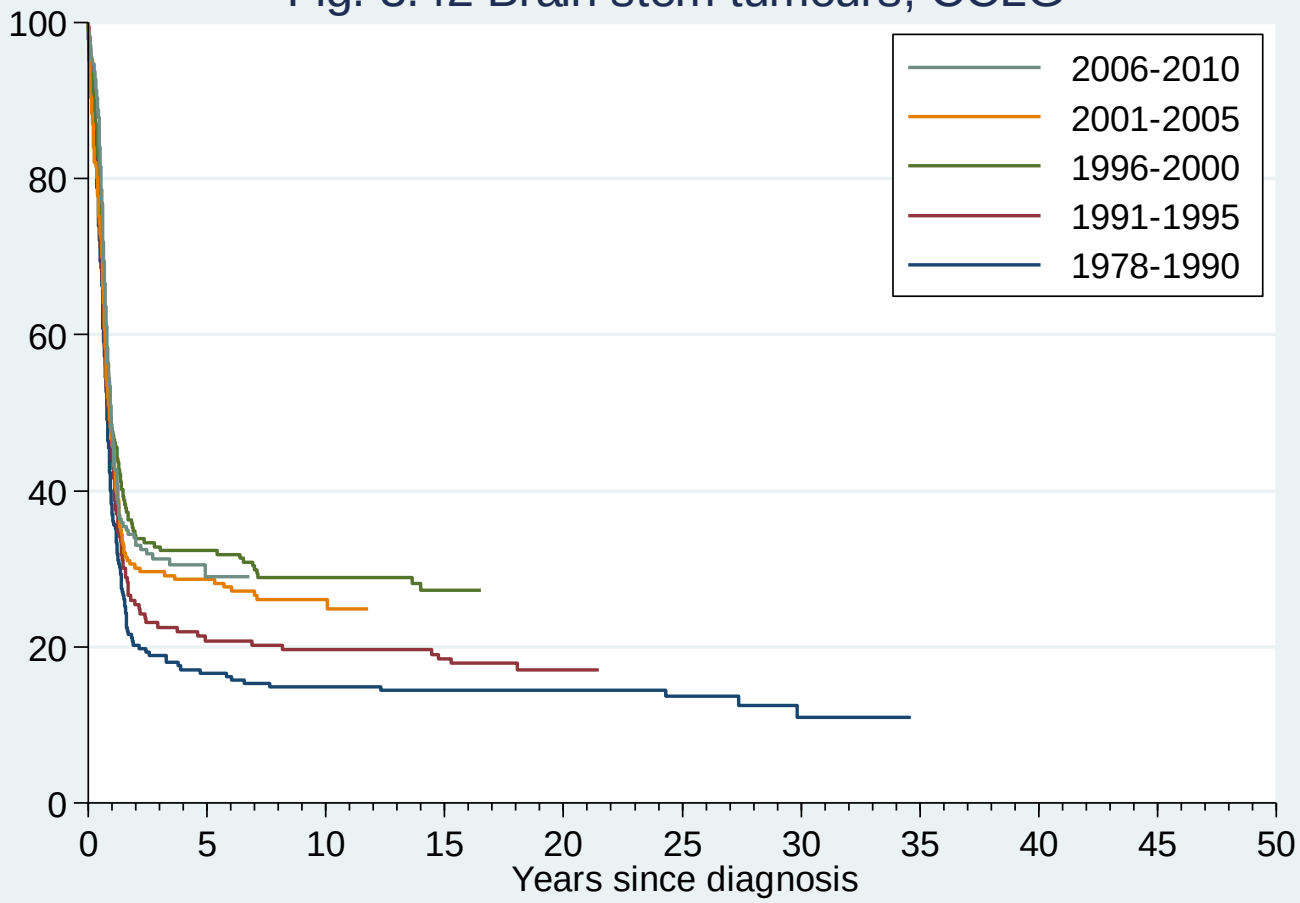


Fig. 3.43 Brain stem astrocytoma, low grade, CCLG

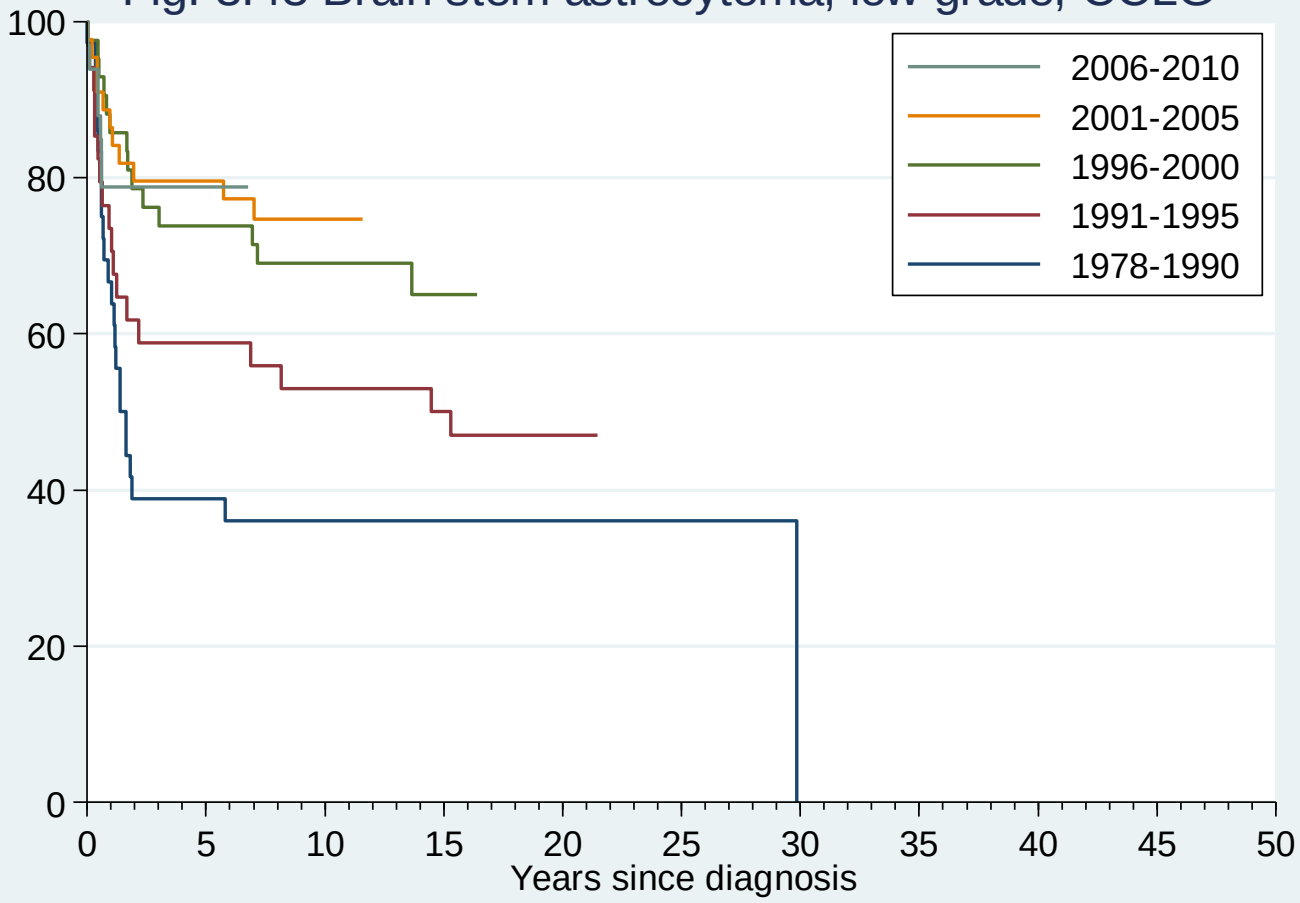


Fig. 3.44 Brain stem astrocytoma, high grade, CCLG

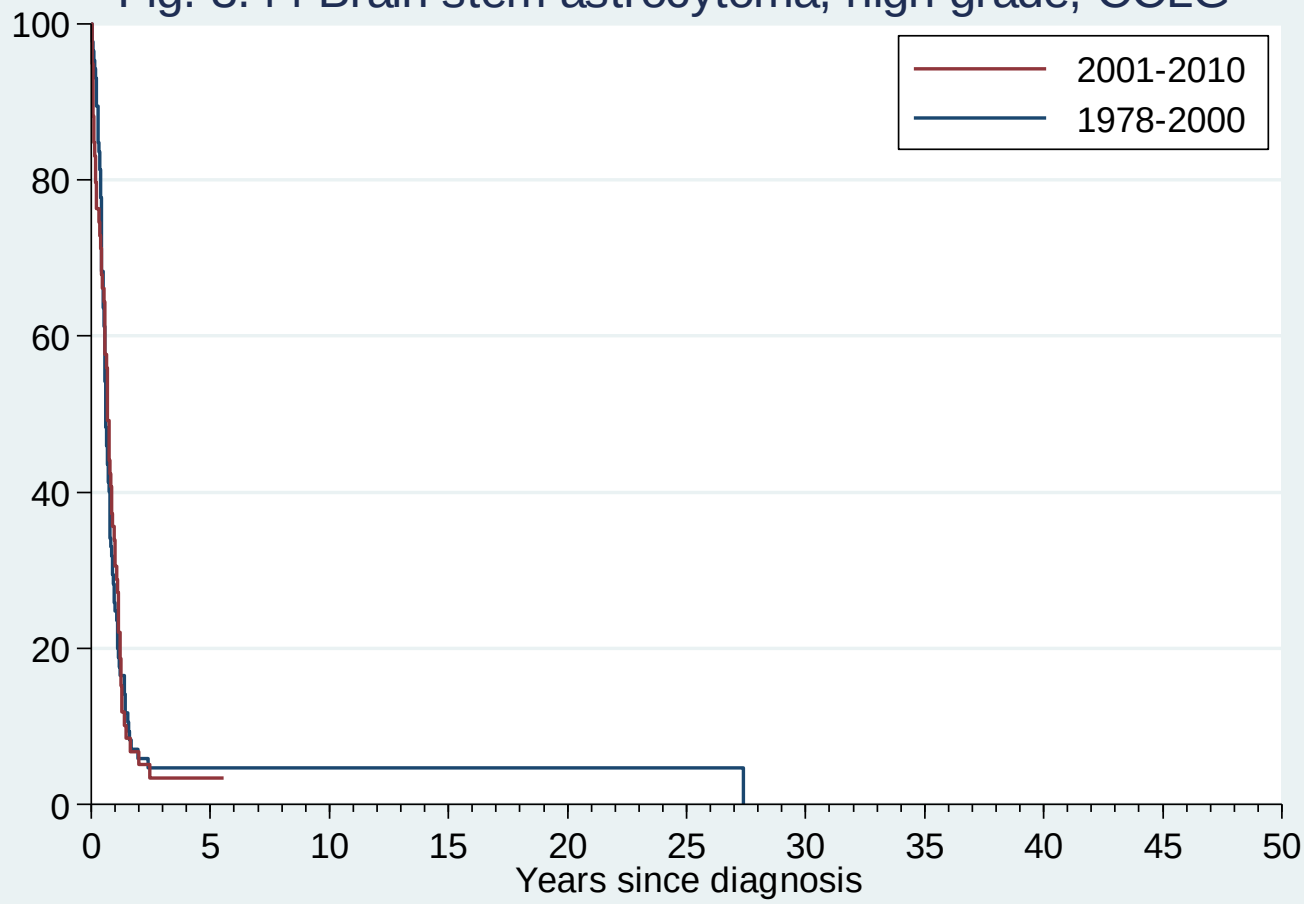


Fig. 3.45 Brain stem tumours - other & unspecified, CCLG

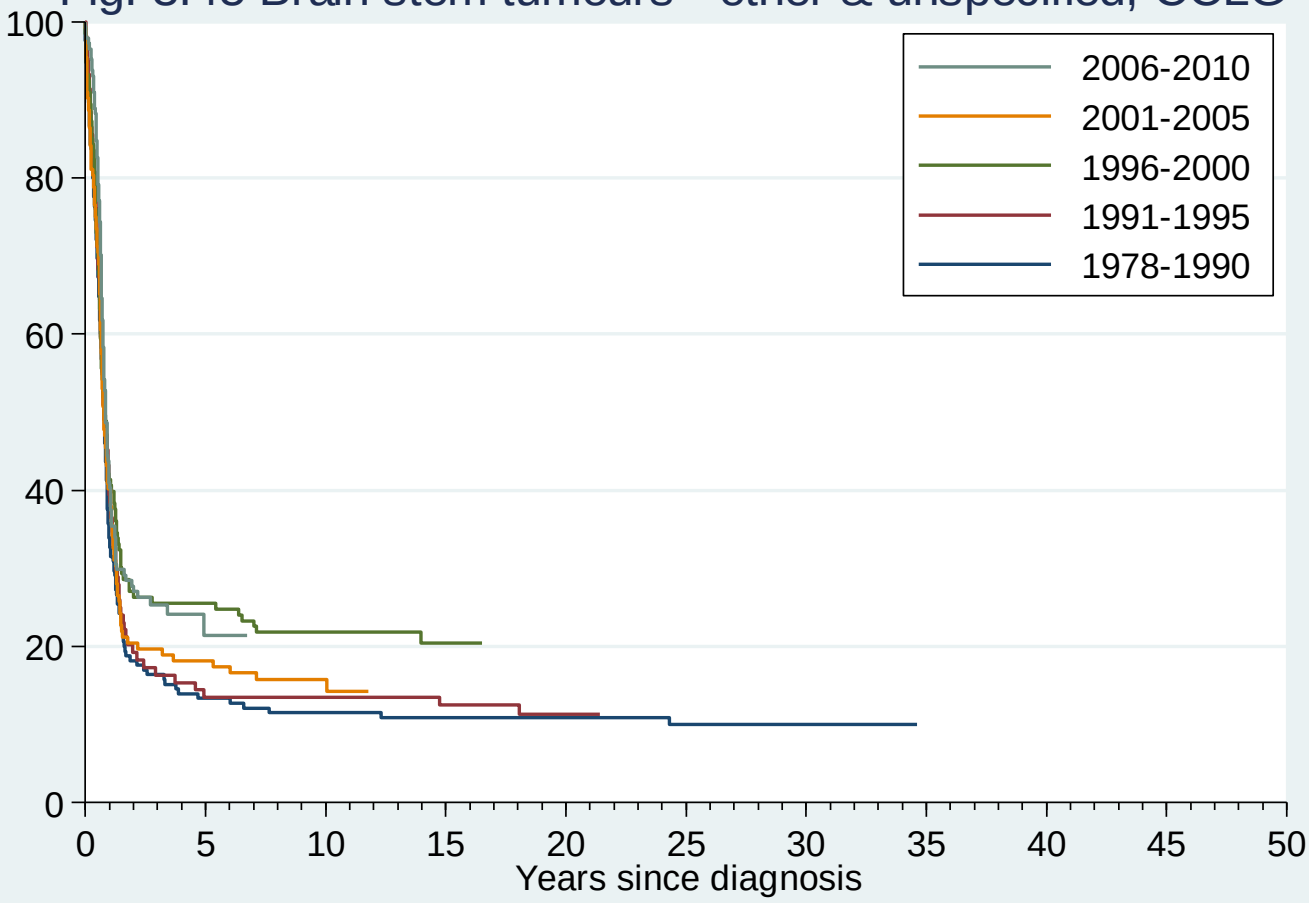


Fig. 3.46 Spinal cord ependymoma, CCLG

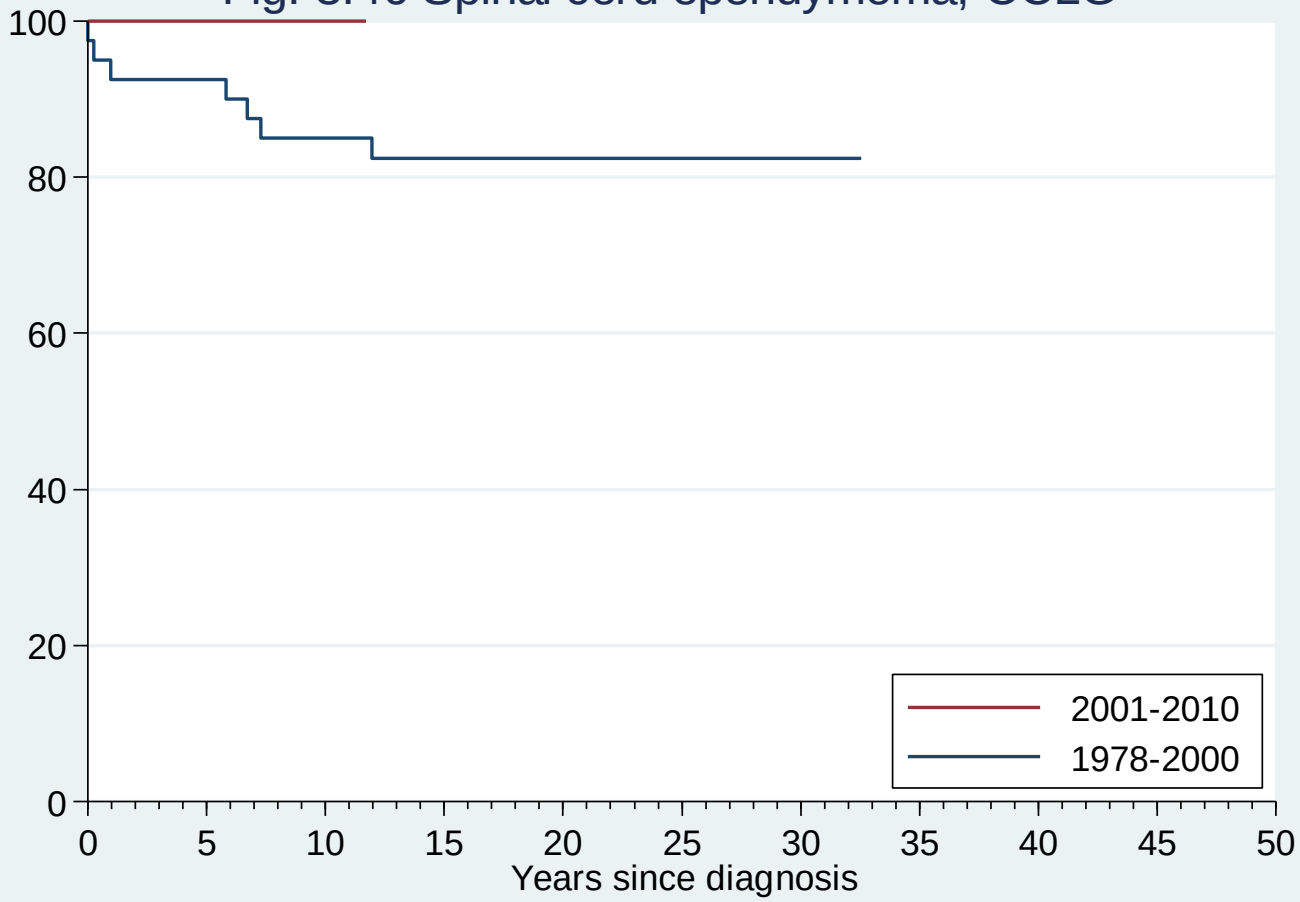


Fig. 3.47 Spinal cord astrocytoma, CCLG

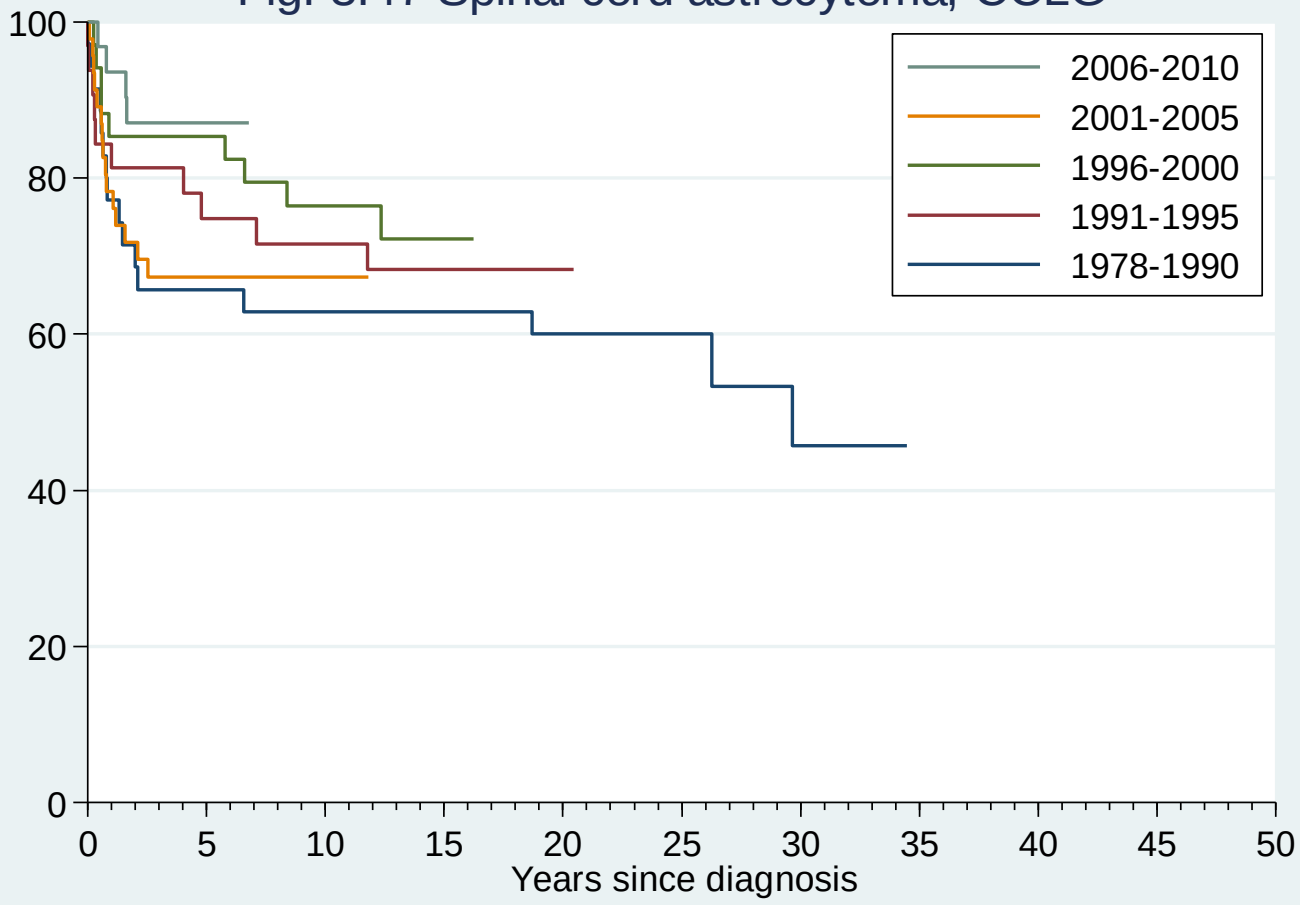


Fig. 3.48 Neuroblastoma, CCLG

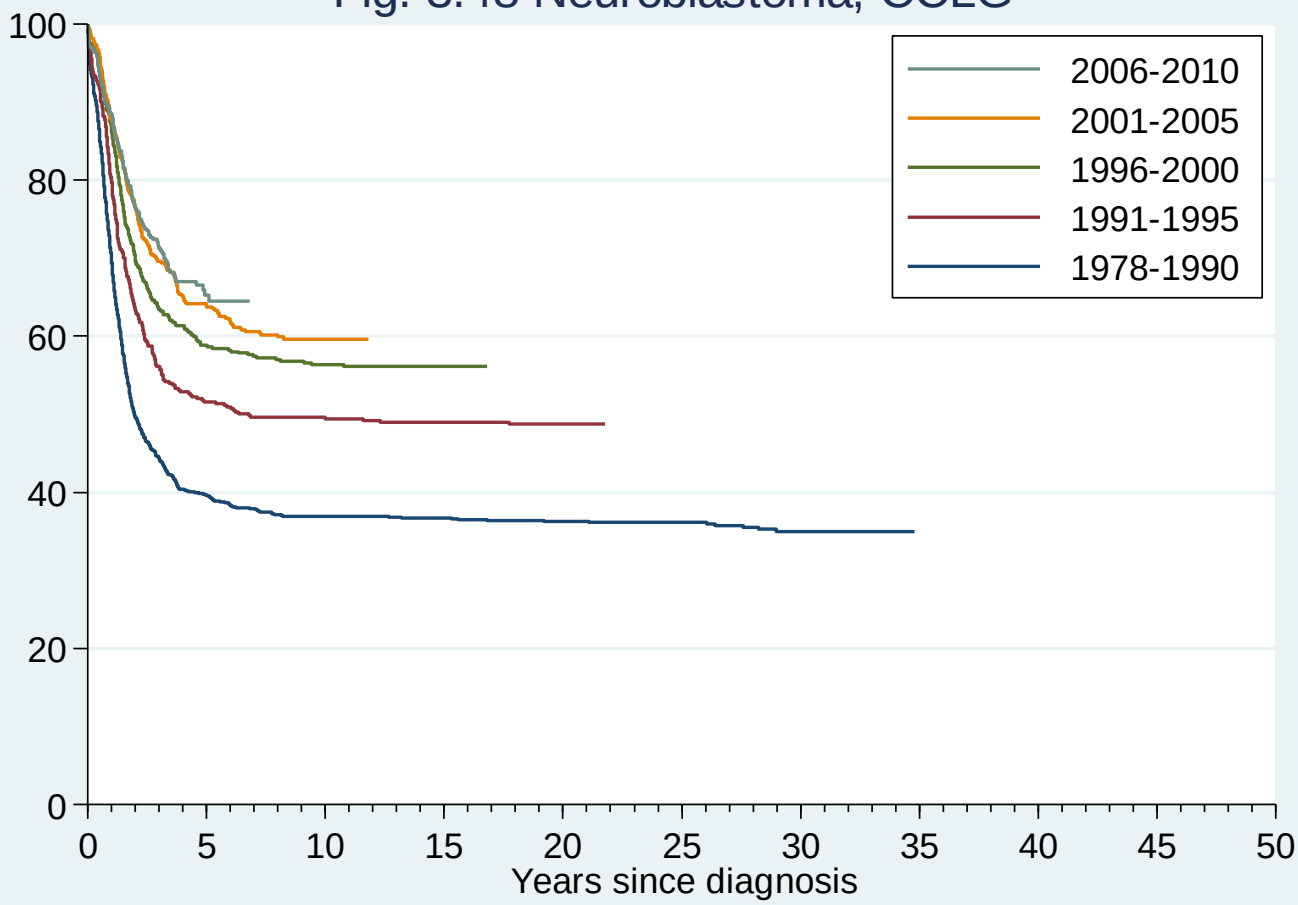


Fig. 3.49 Retinoblastoma, bilateral, CCLG

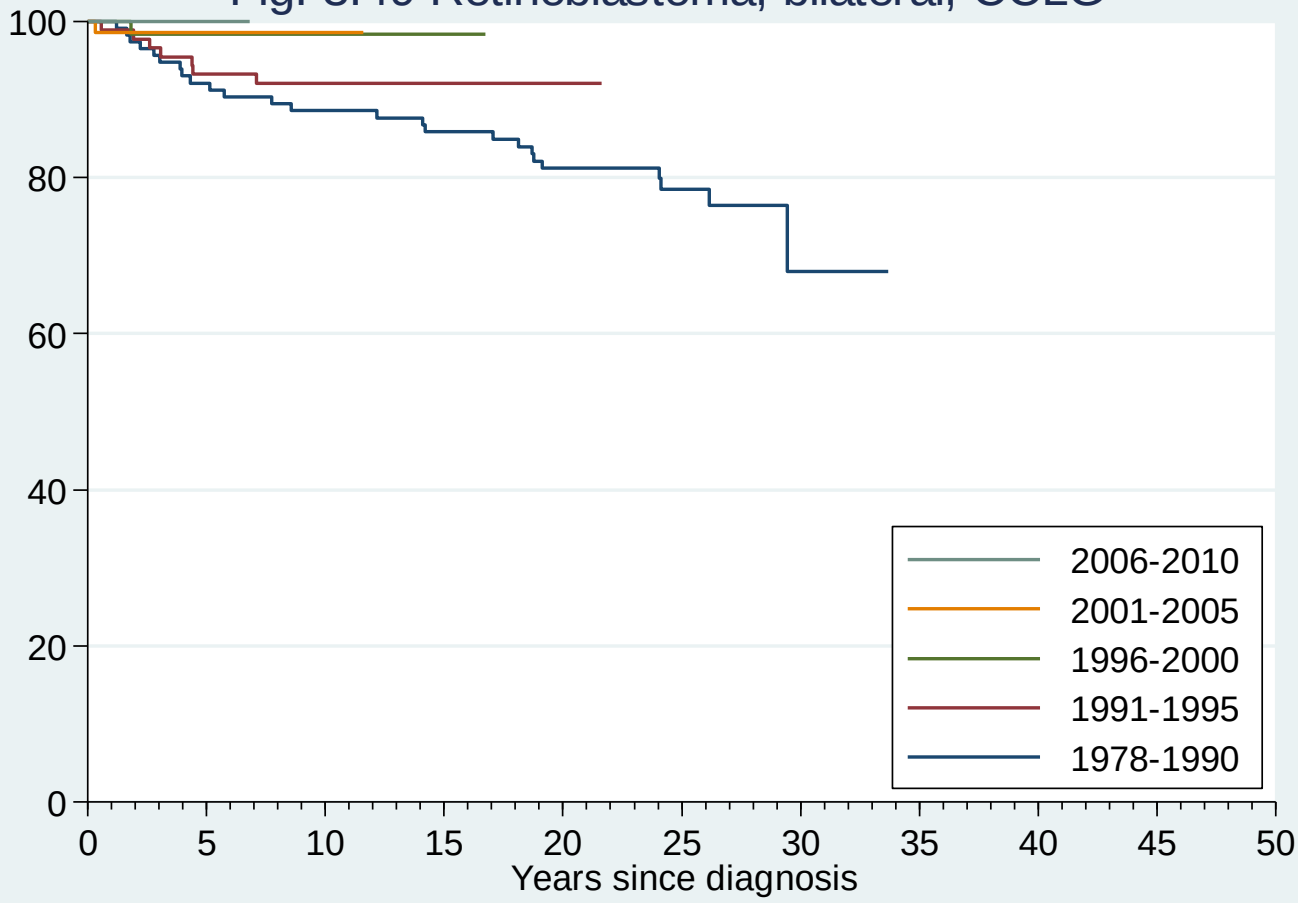


Fig. 3.50 Retinoblastoma, unilateral, CCLG

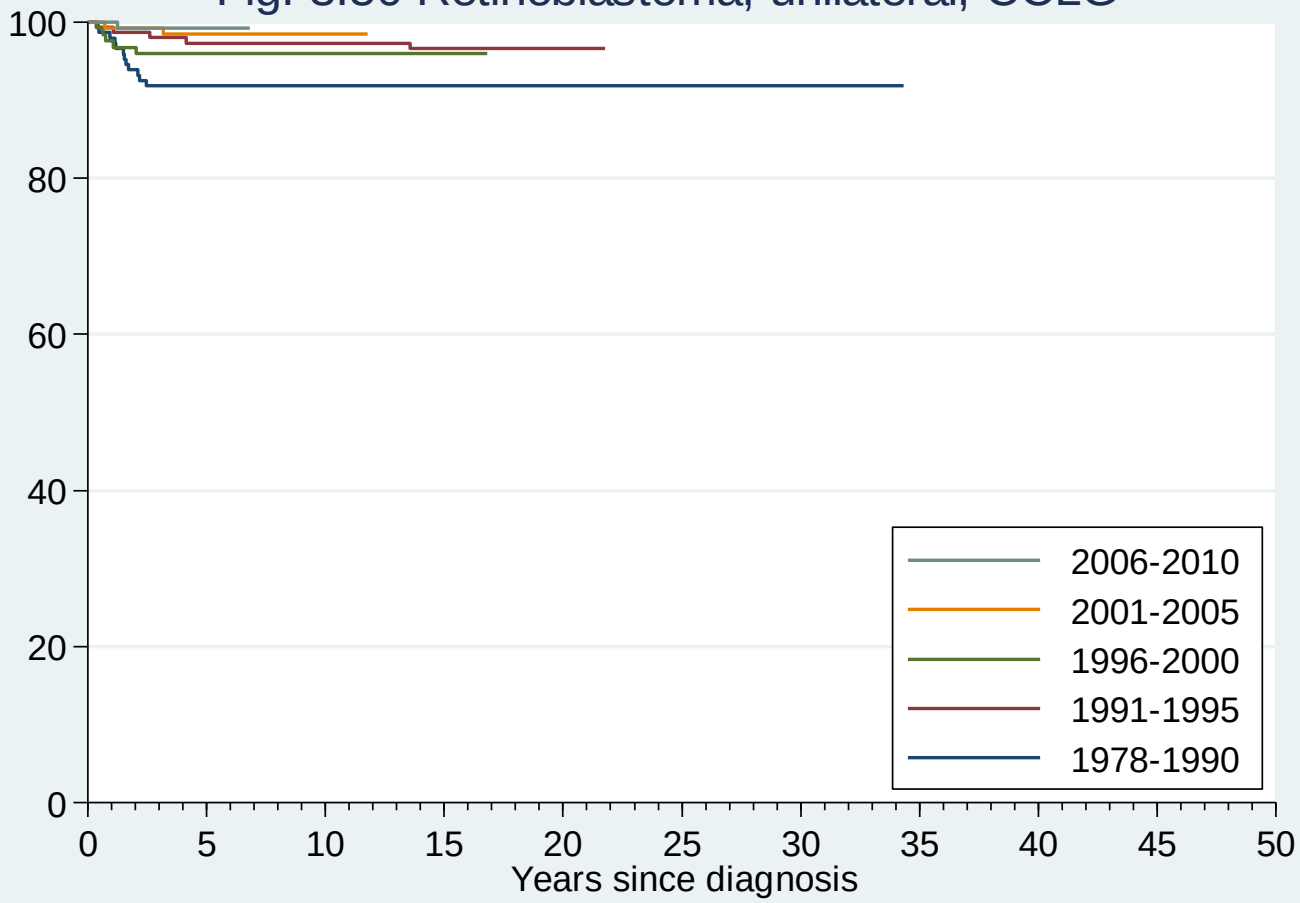


Fig. 3.51 Wilms tumour, CCLG

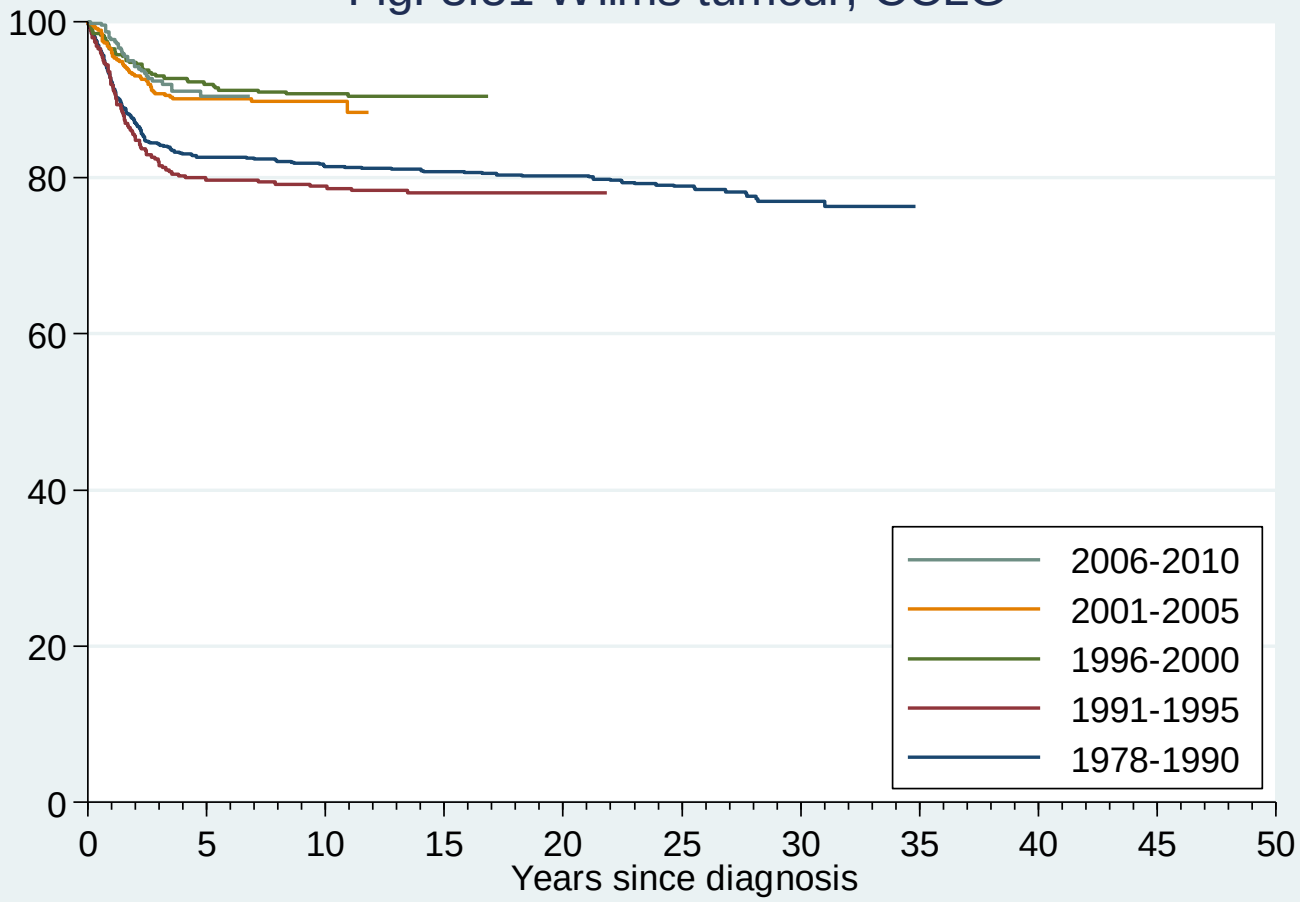


Fig. 3.52 Rhabdoid renal tumour, CCLG

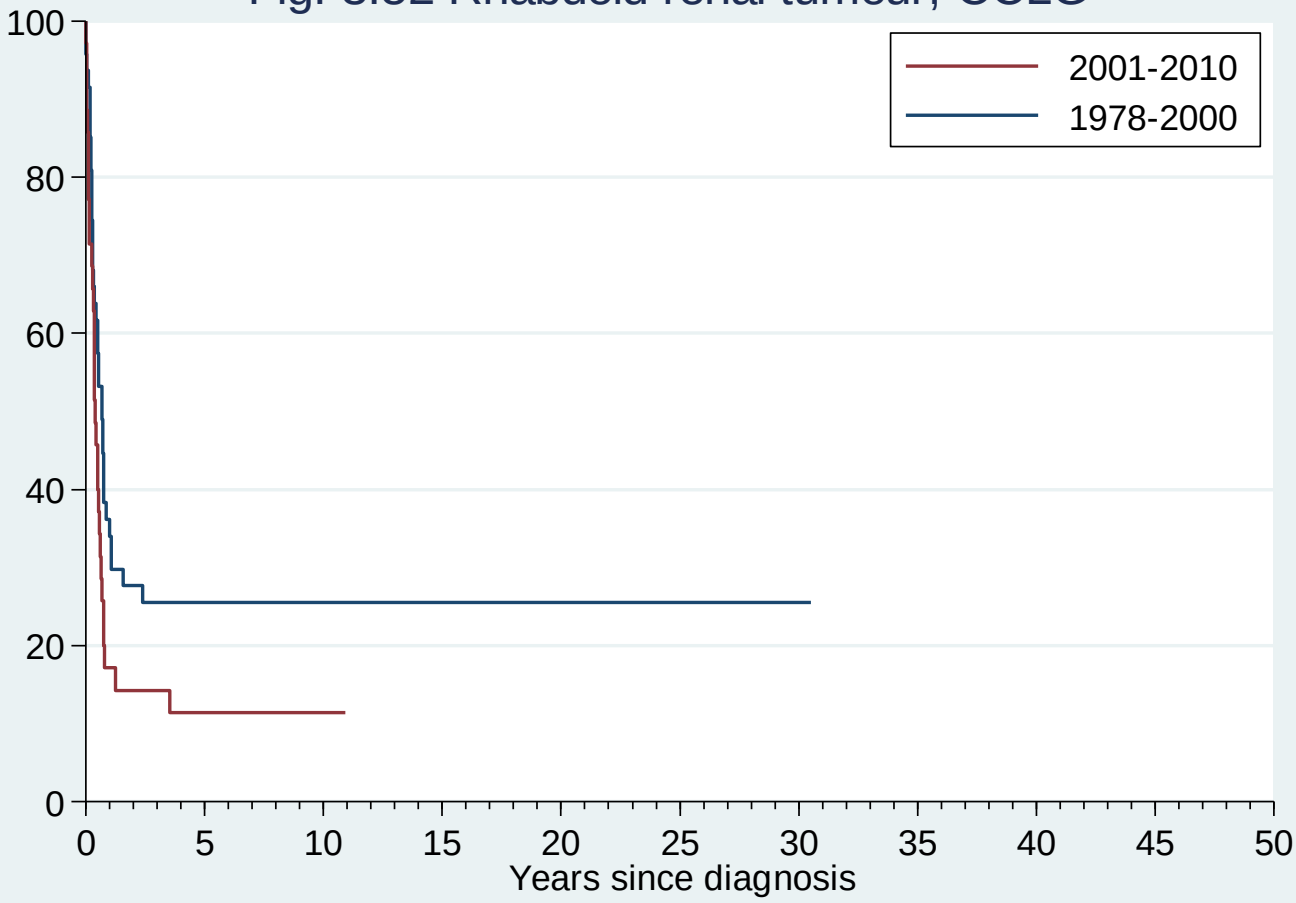


Fig. 3.53 Renal clear-cell sarcoma, CCLG

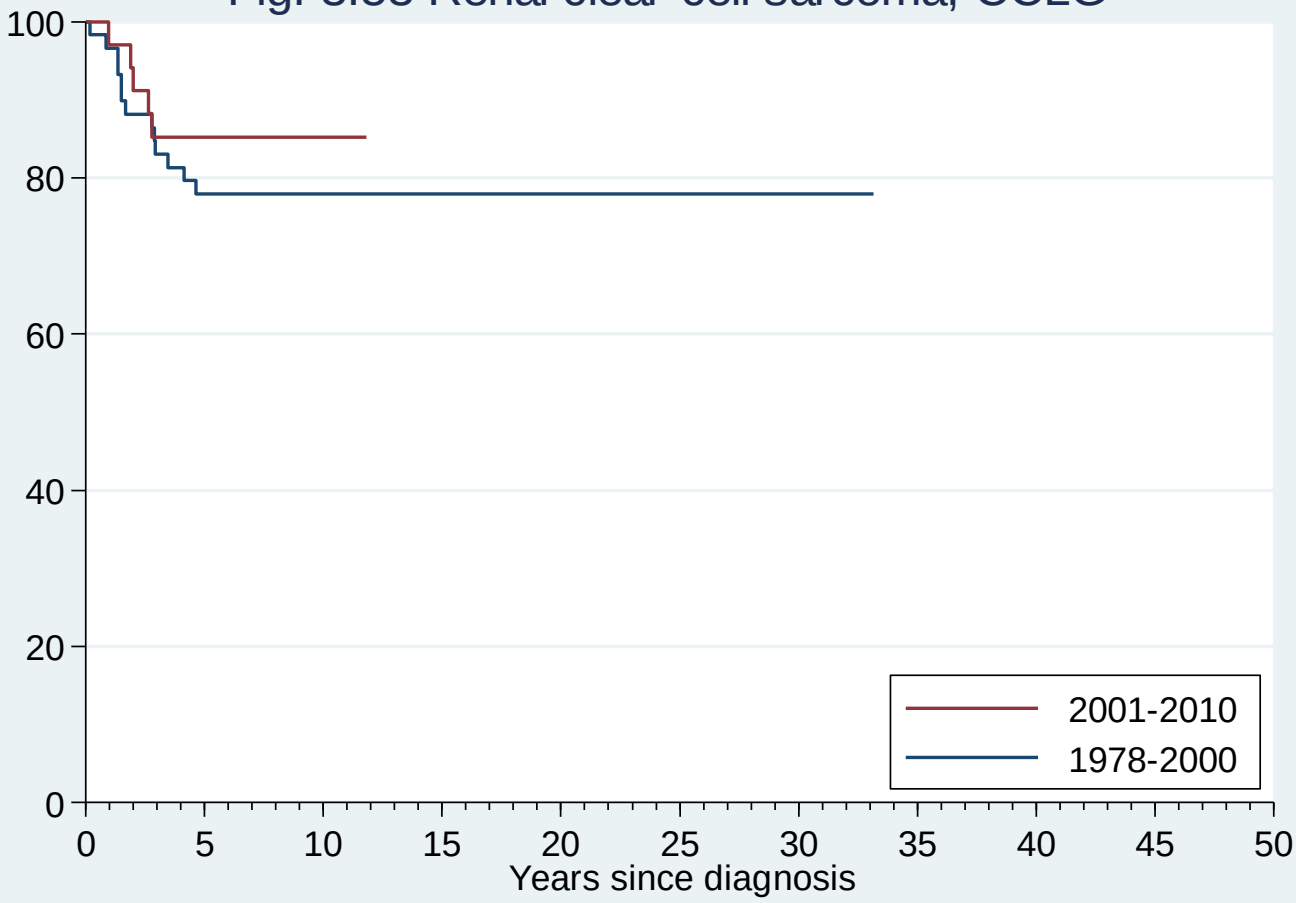


Fig. 3.54 Renal PNET, CCLG

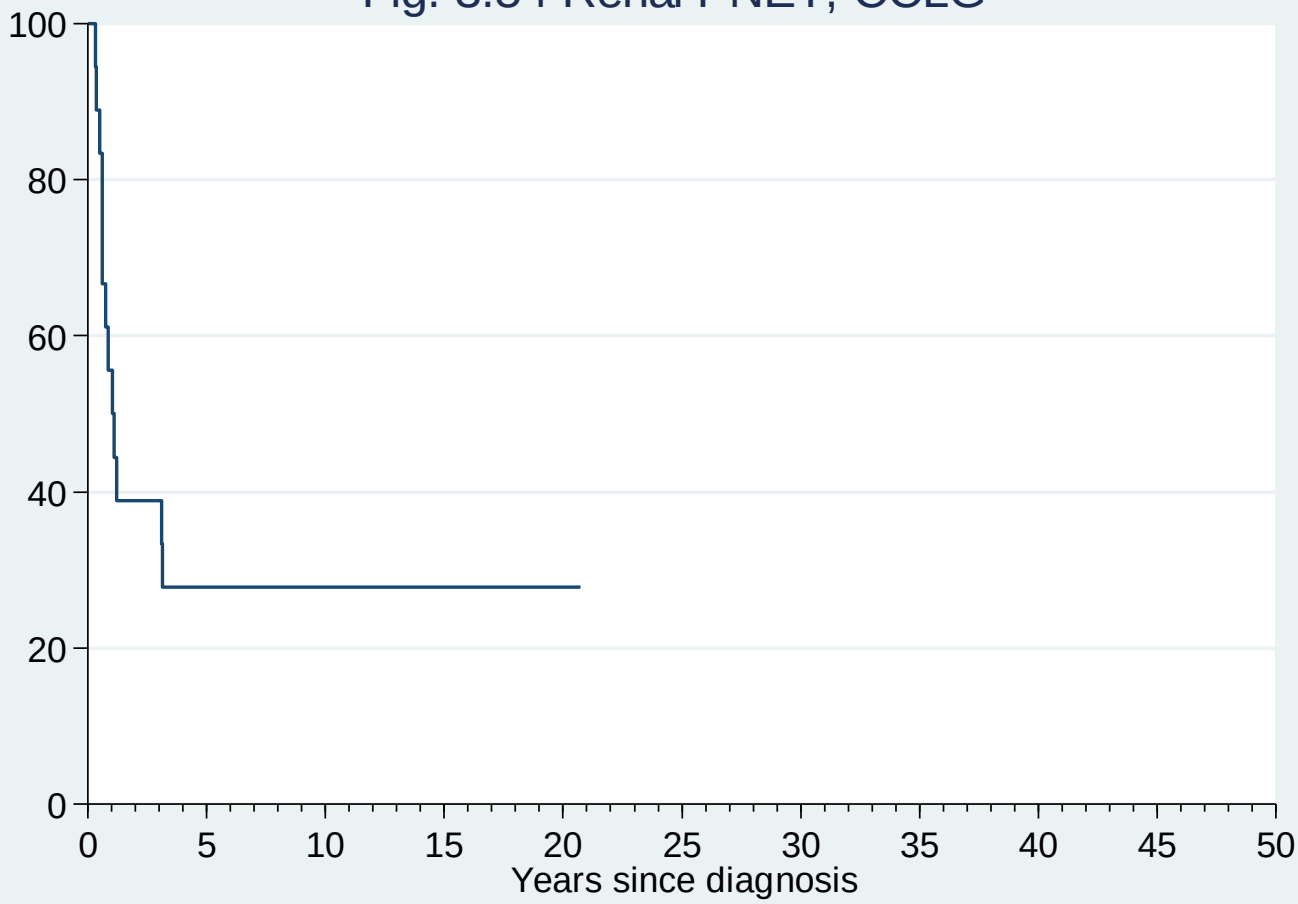


Fig. 3.55 Renal carcinoma, CCLG

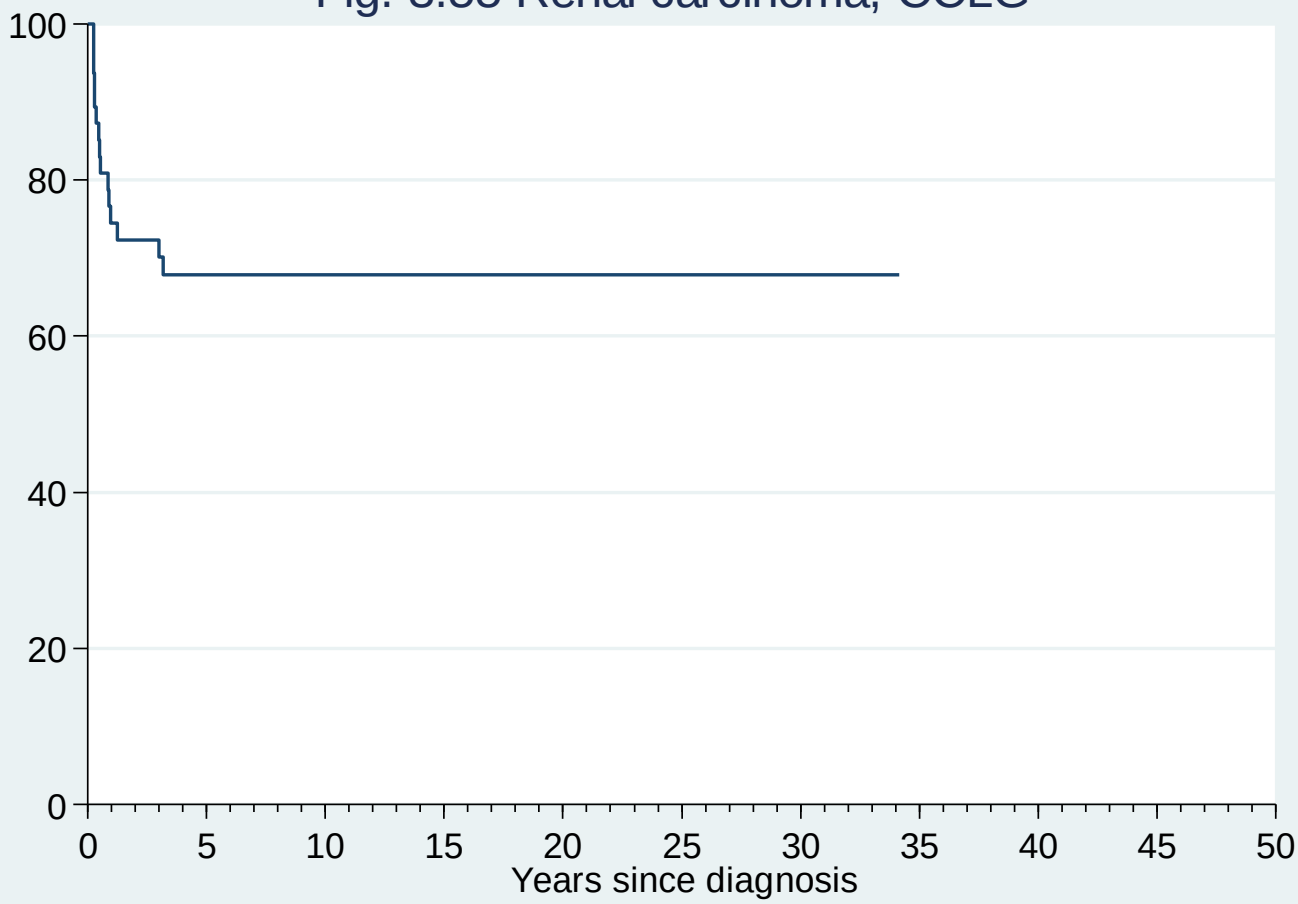


Fig. 3.56 Hepatoblastoma, CCLG

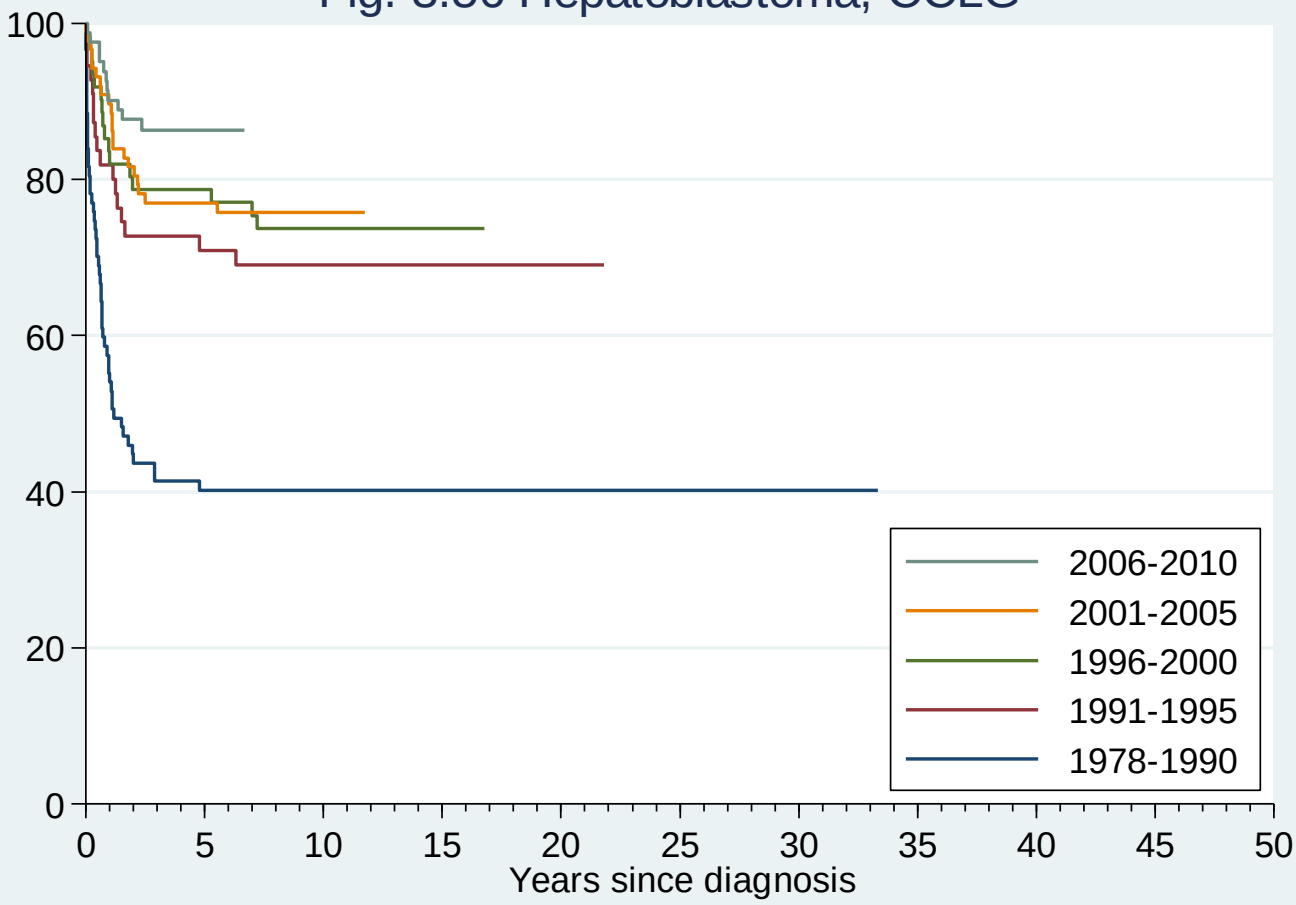


Fig. 3.57 Hepatic carcinoma, CCLG

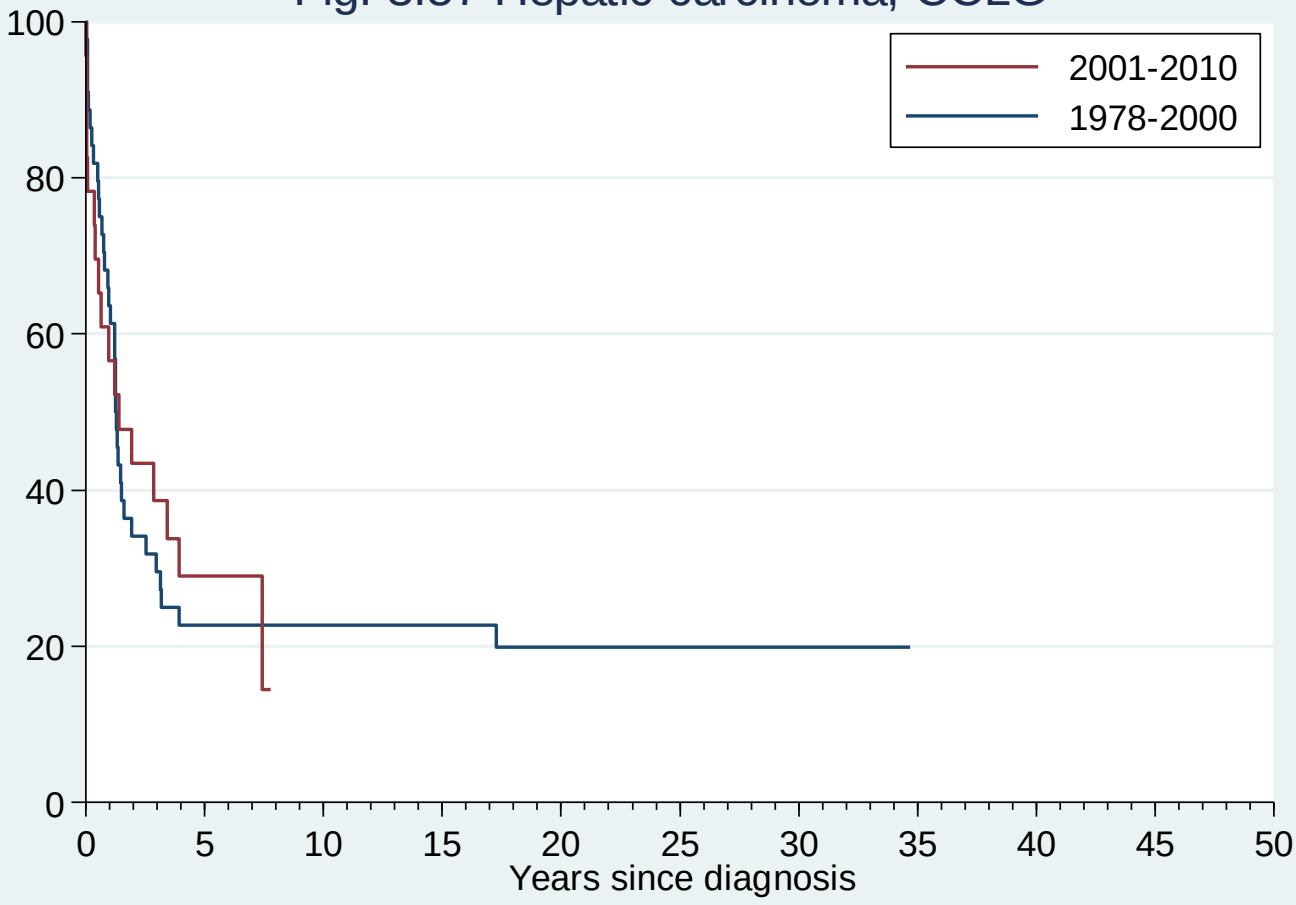


Fig. 3.58 Osteosarcoma, CCLG

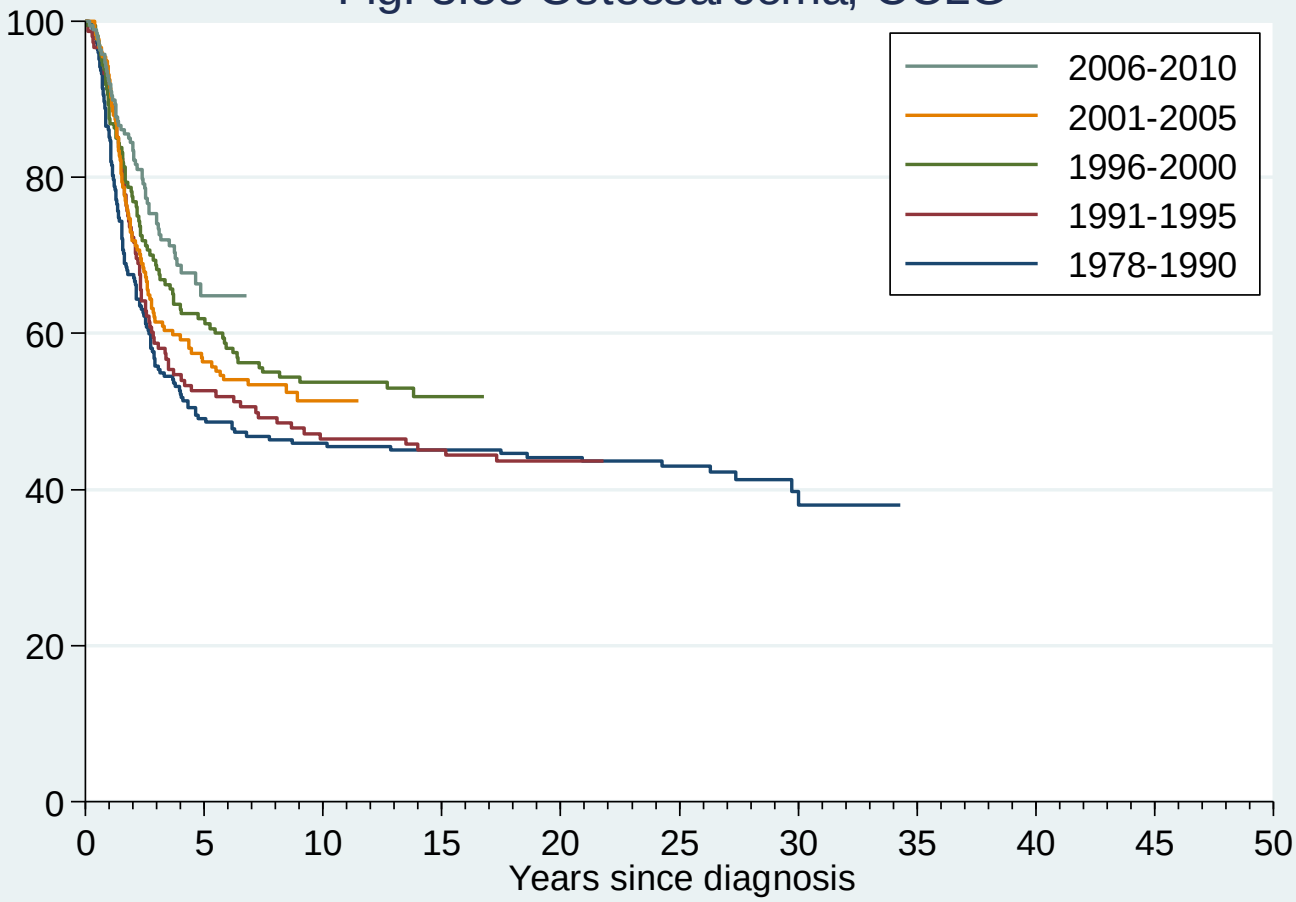


Fig. 3.59 Chondrosarcoma, CCLG

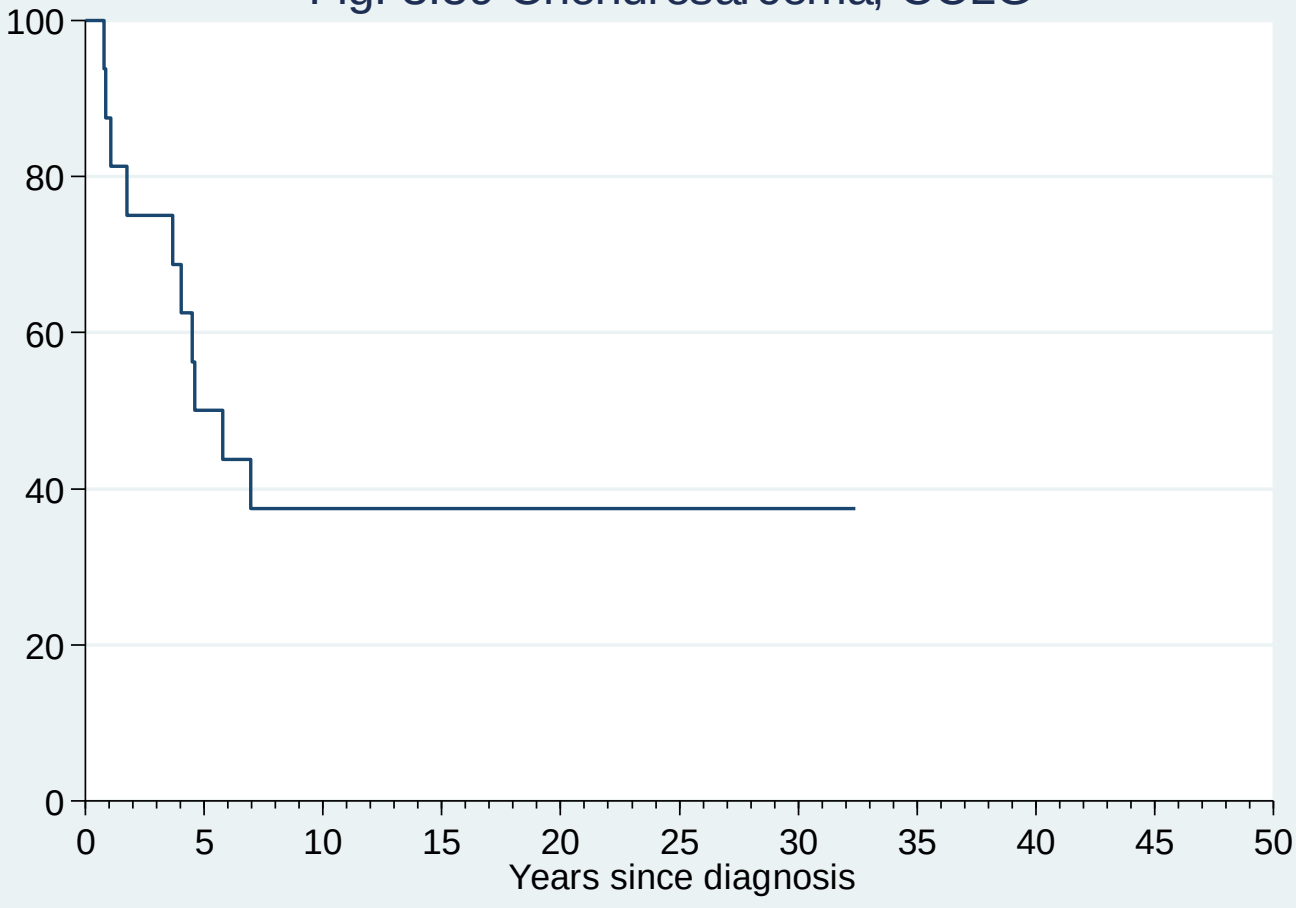


Fig. 3.60 Ewing sarcoma of bone, CCLG

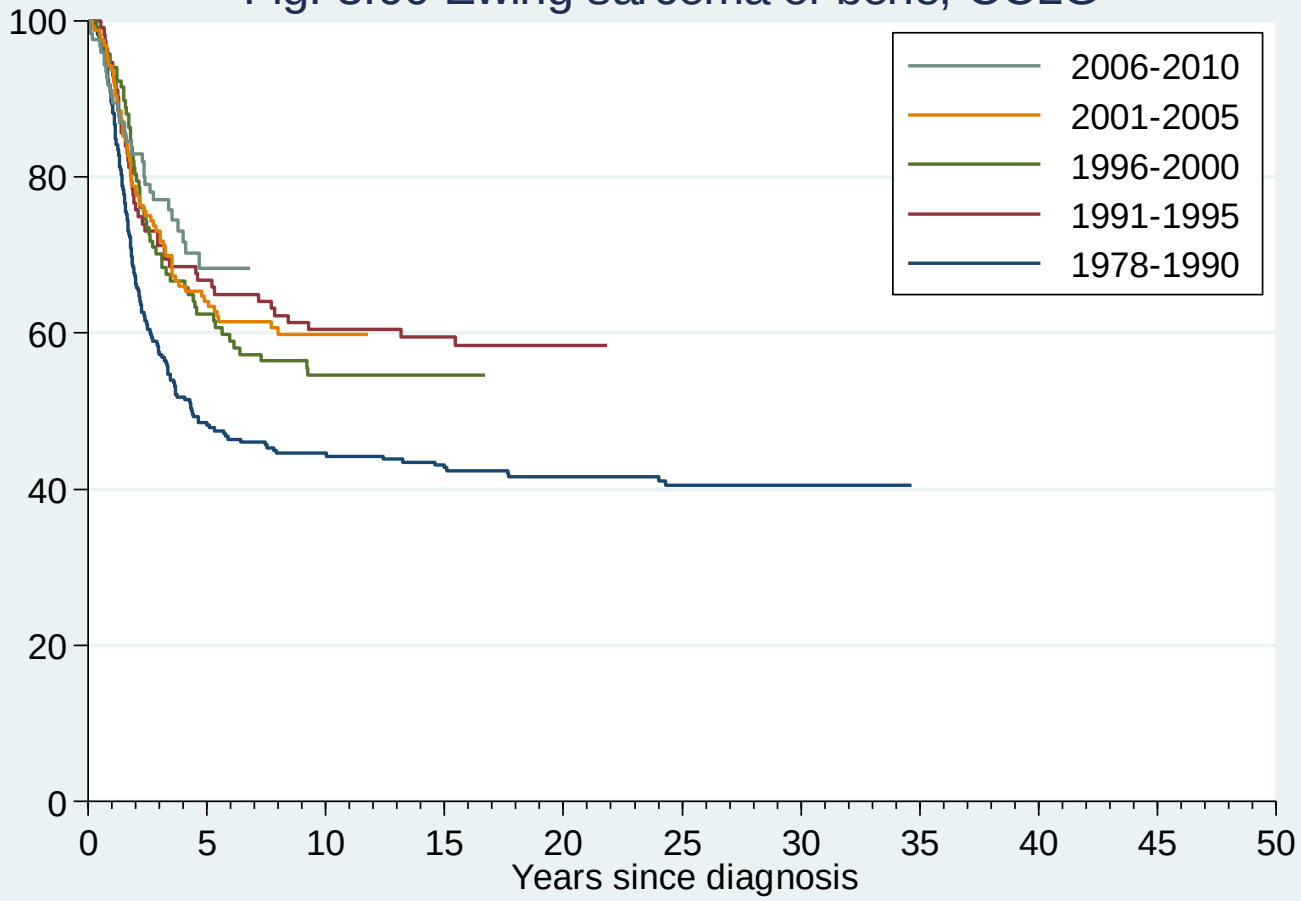


Fig. 3.61 Rhabdomyosarcoma, CCLG

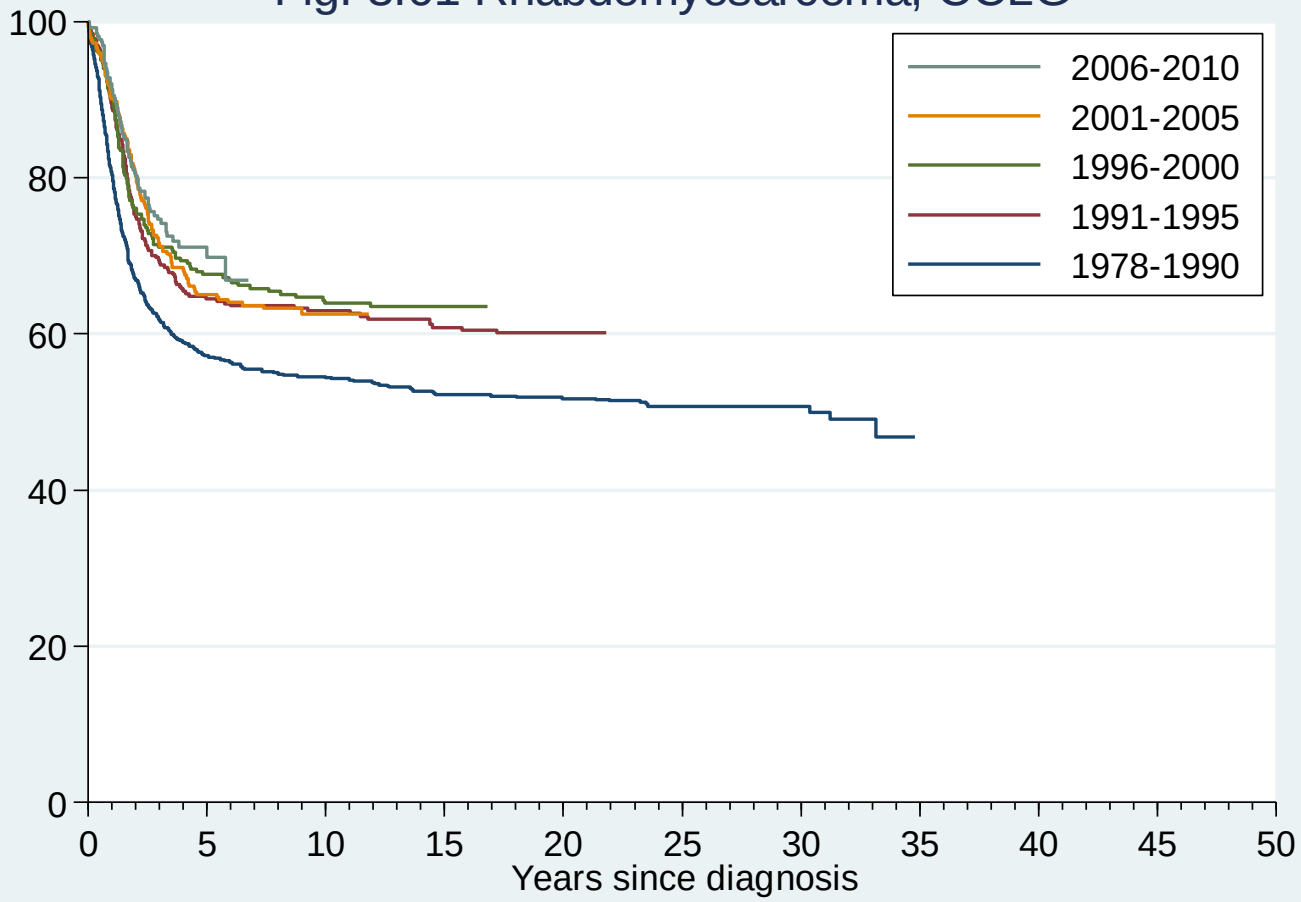


Fig. 3.62 Malignant peripheral nerve sheath tumour, CCLG

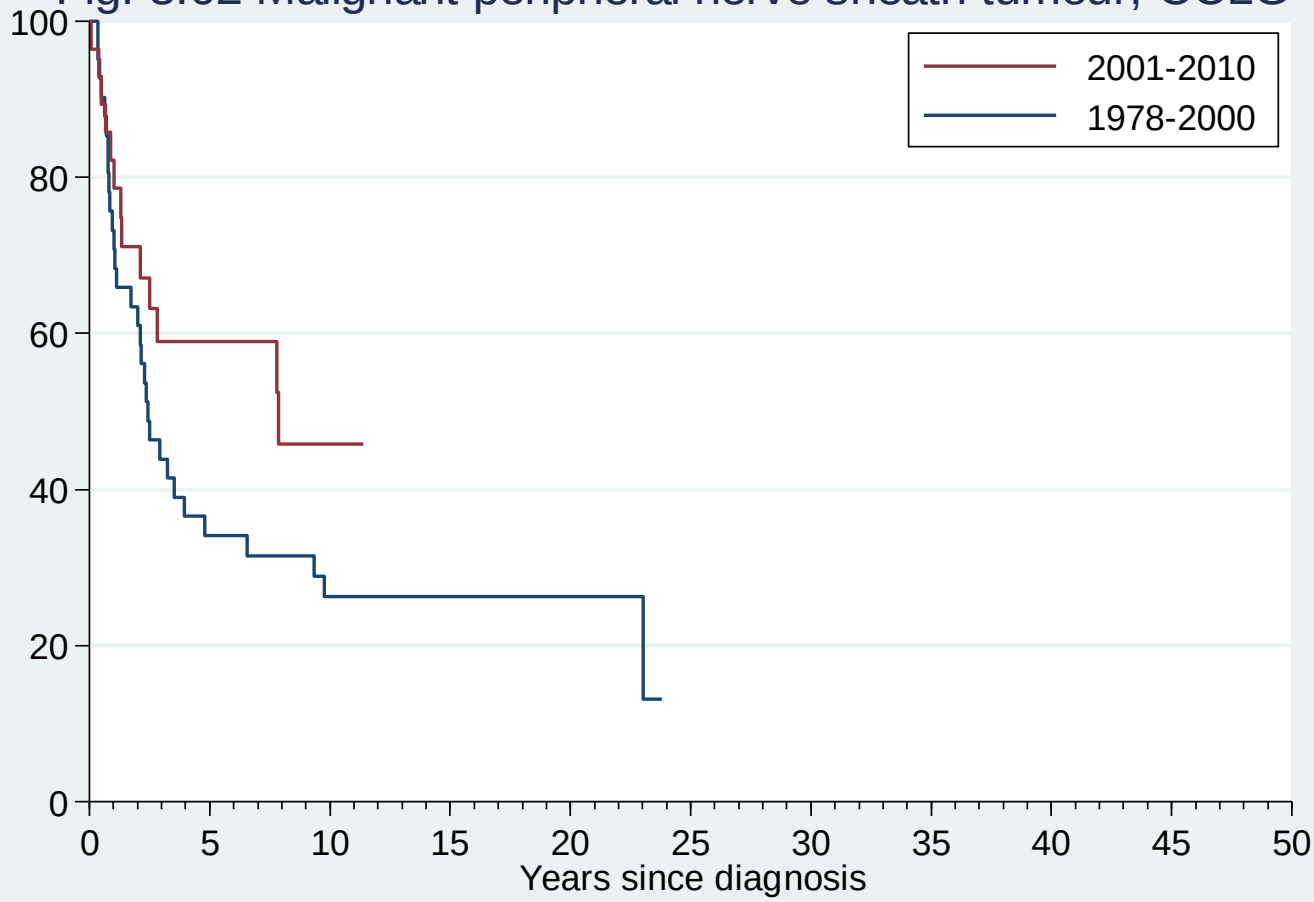


Fig. 3.63 Other fibrosarcoma etc, CCLG

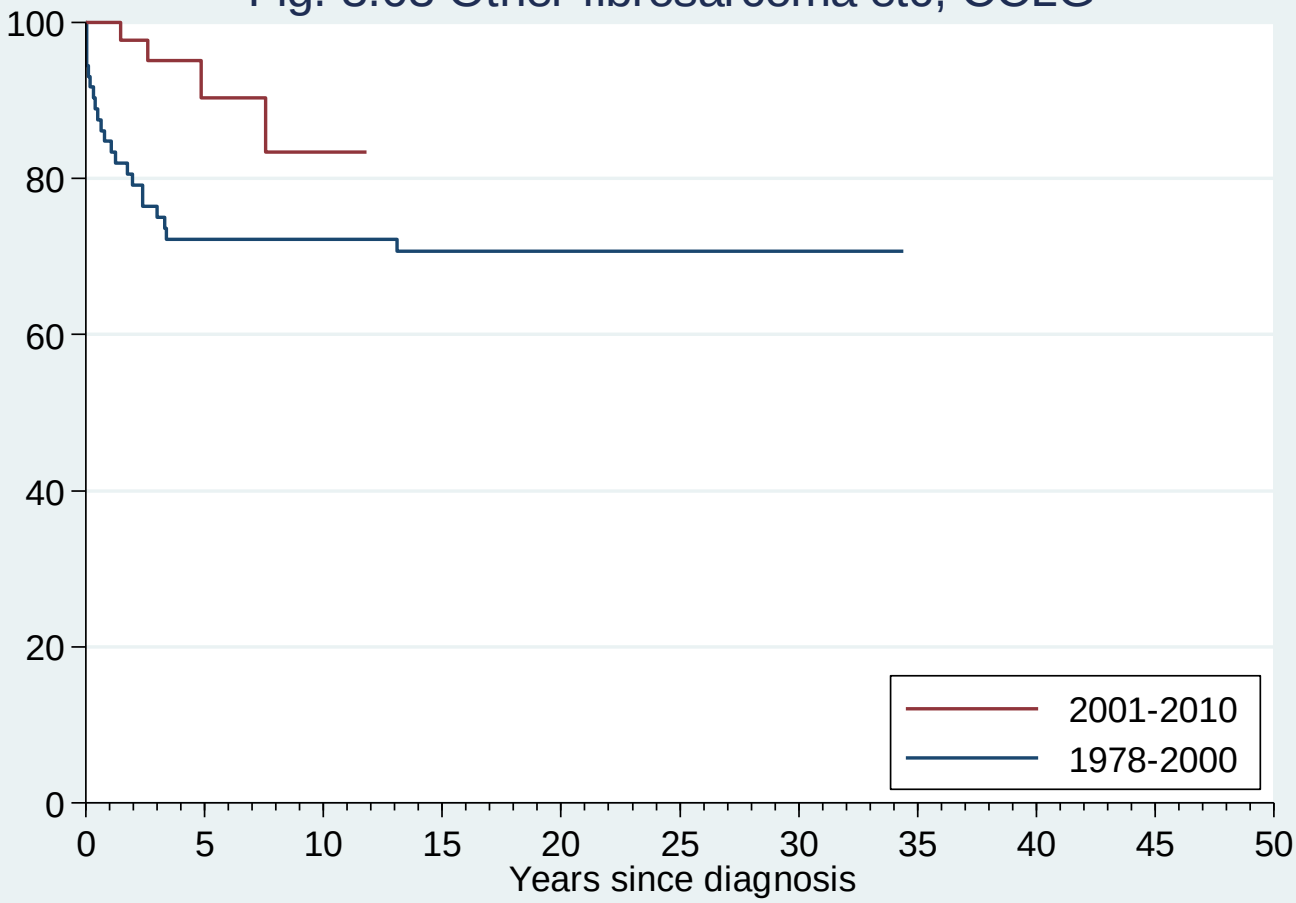


Fig. 3.64 Extraosseous ESFT, CCLG

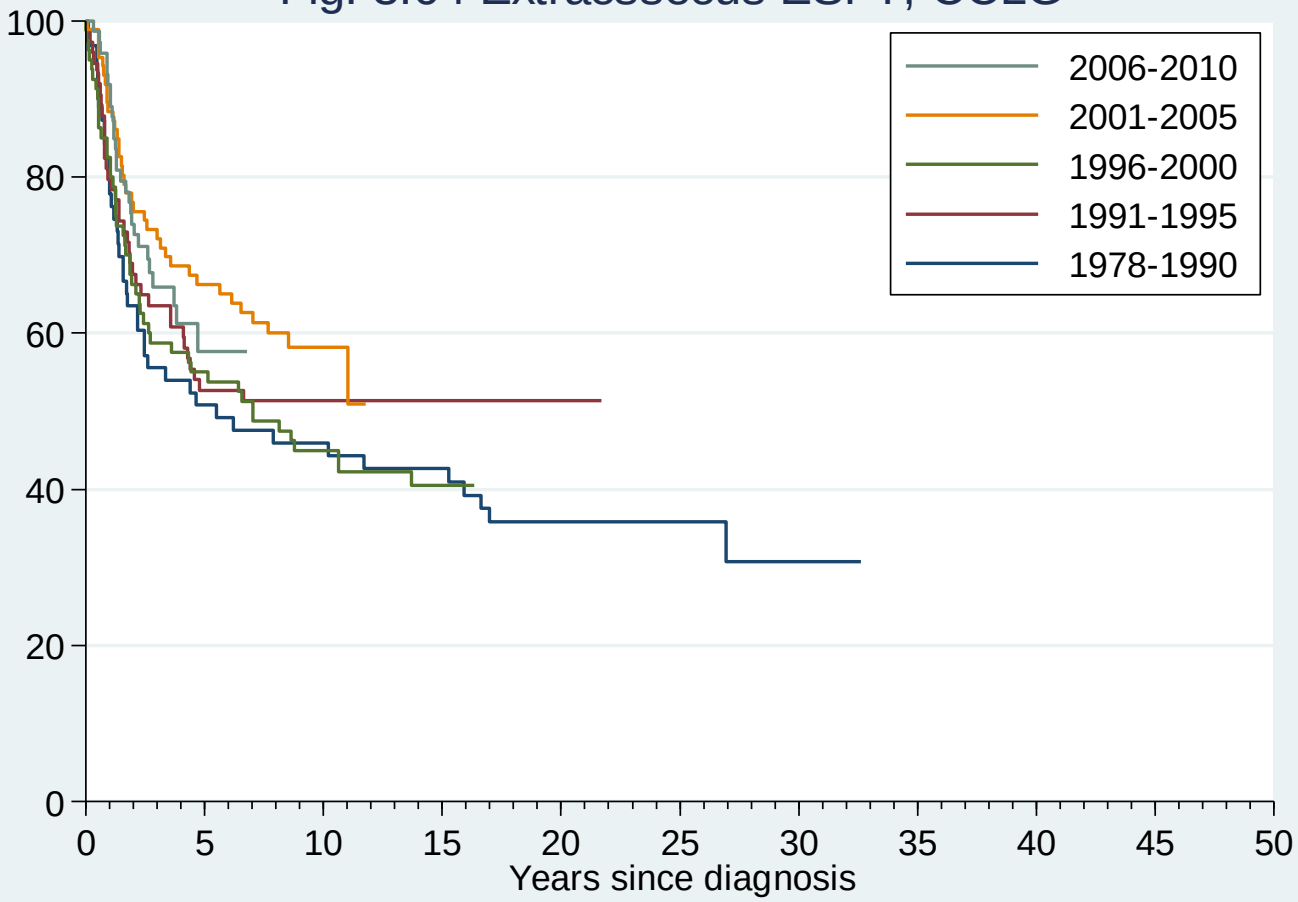


Fig. 3.65 Extrarenal rhabdoid tumour, CCLG

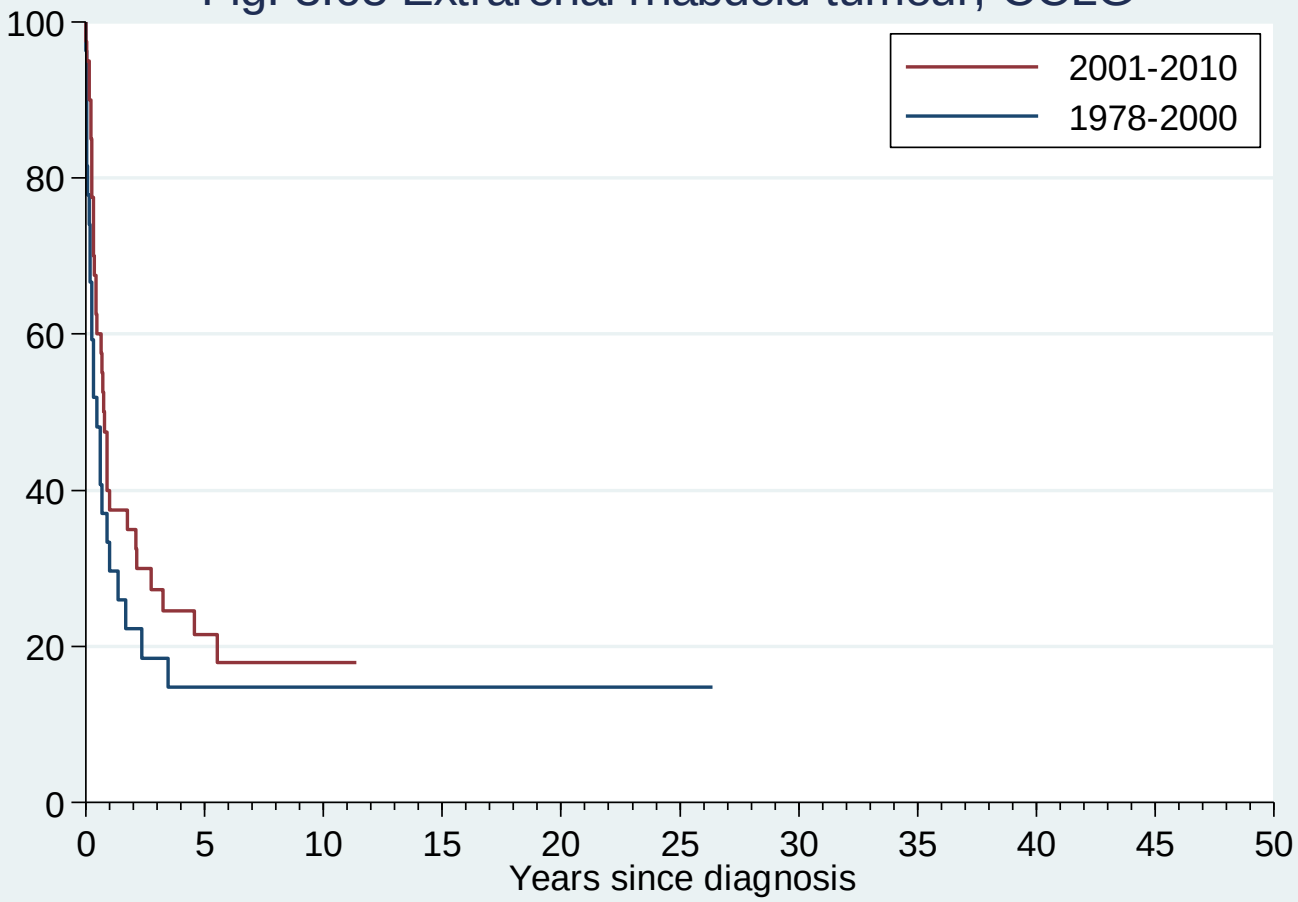


Fig. 3.66 Synovial sarcoma, CCLG

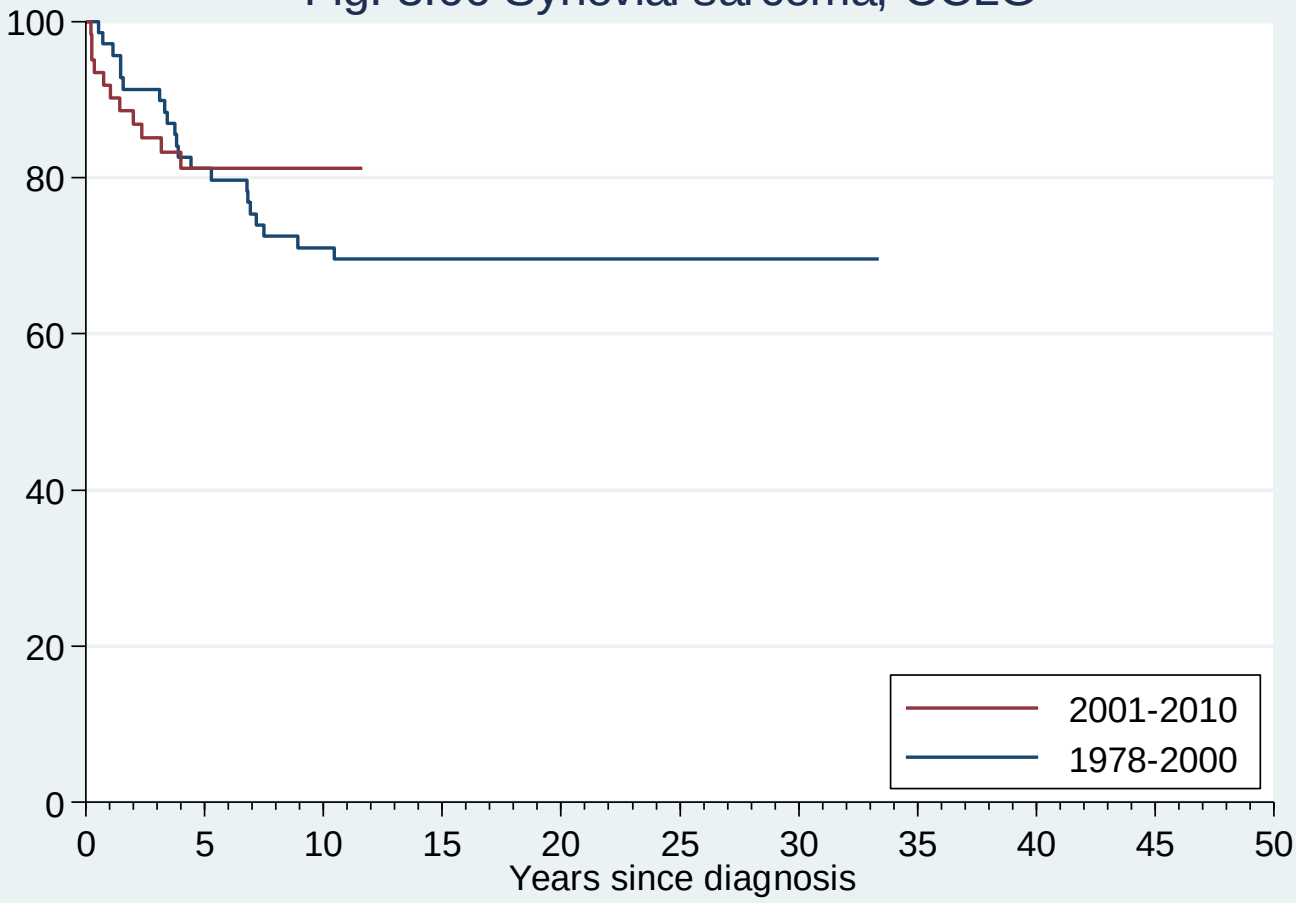


Fig. 3.67 Leiomyosarcoma, CCLG

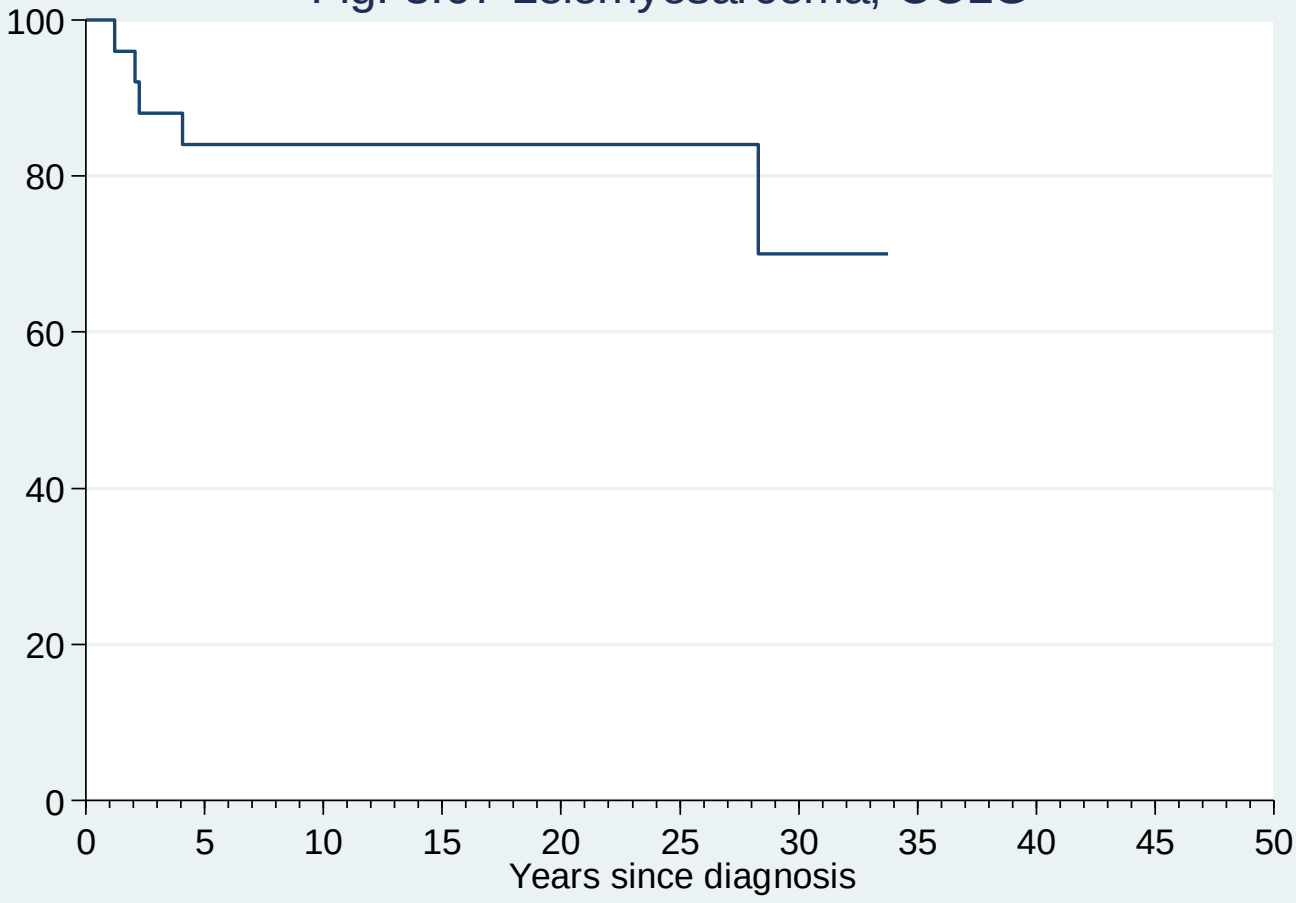


Fig. 3.68 Fibrohistiocytic tumours, CCLG

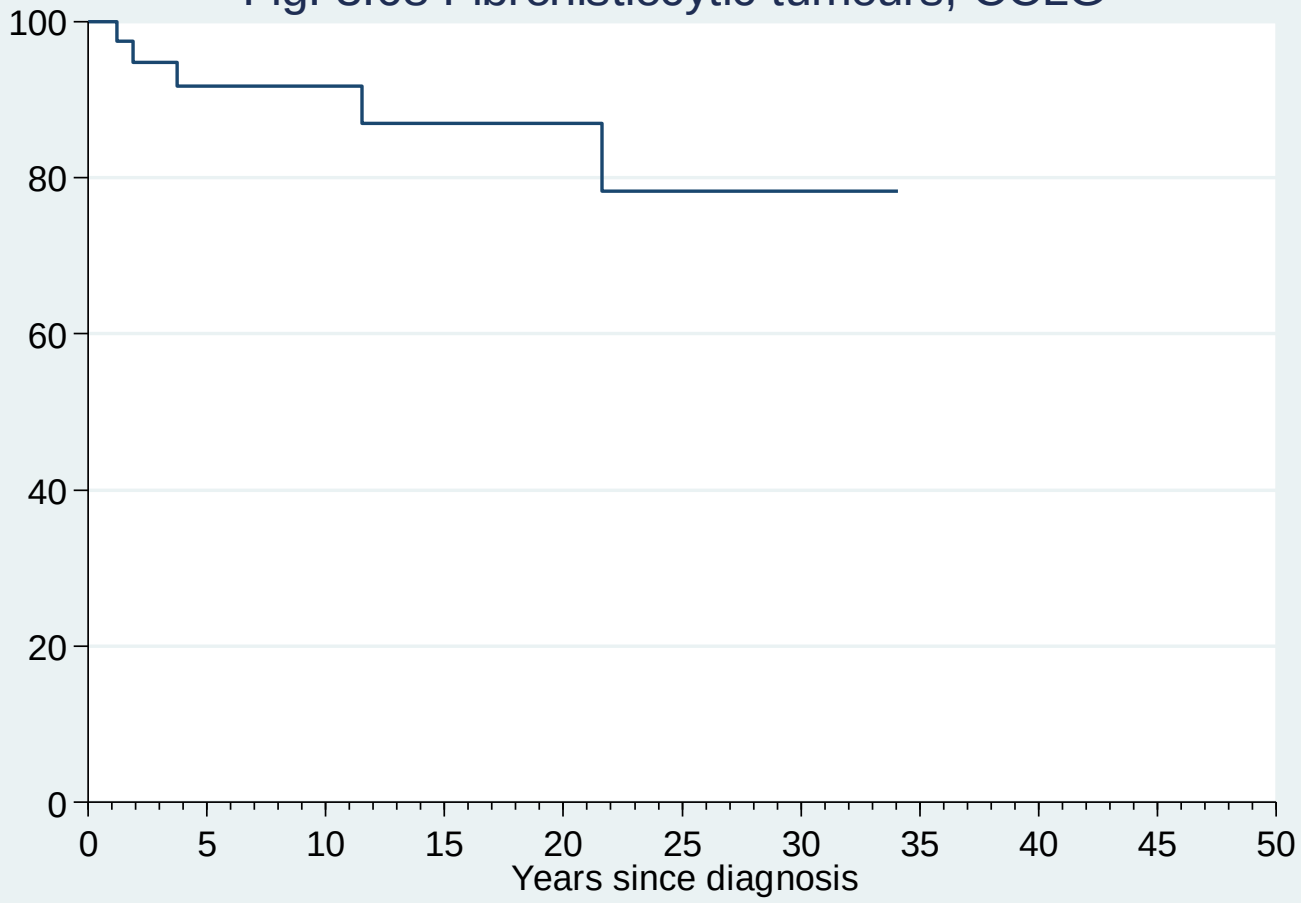


Fig. 3.69 Alveolar soft part sarcoma, CCLG

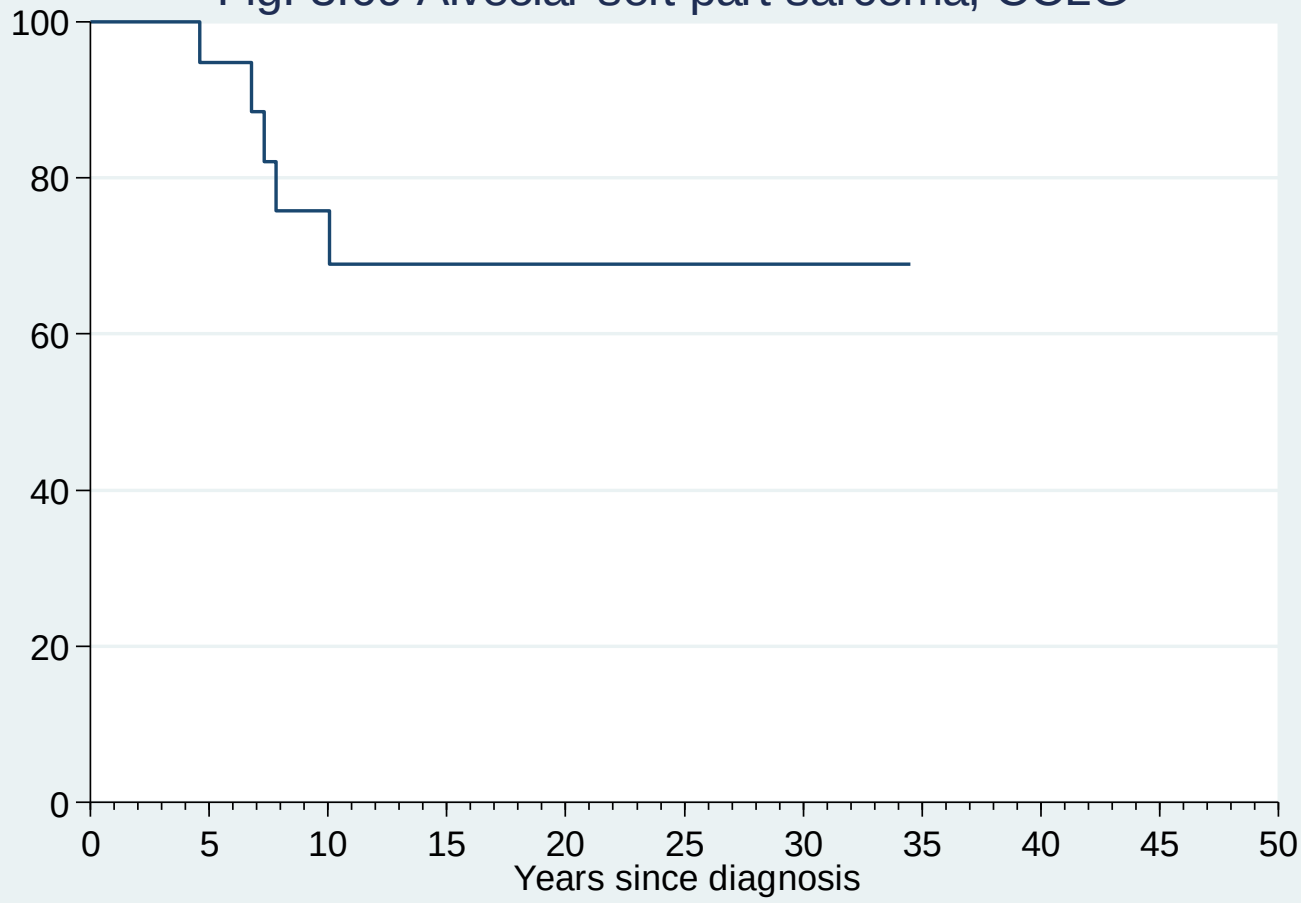


Fig. 3.70 Desmoplastic small round cell tumour, CCLG

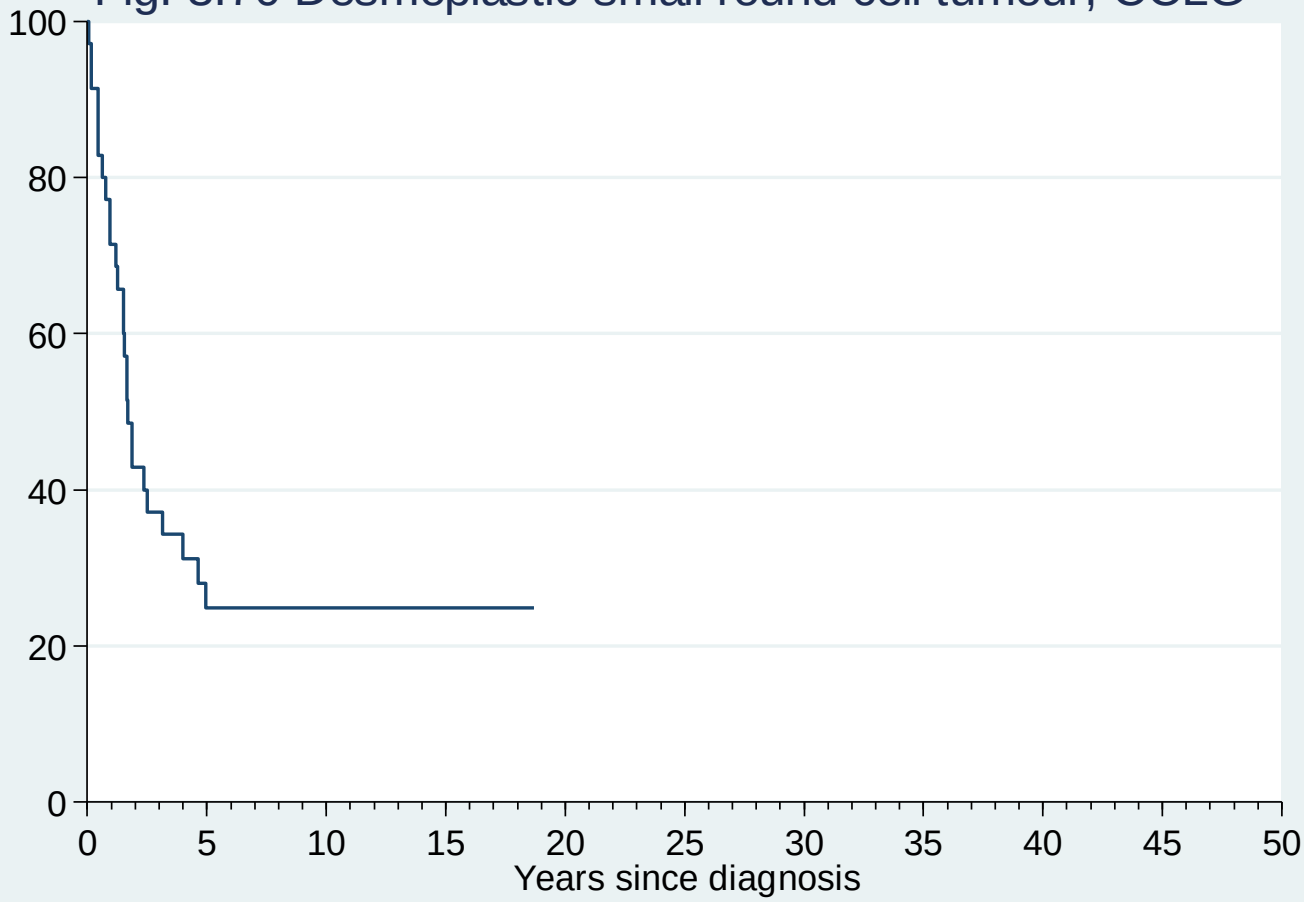


Fig. 3.71 Other specified soft-tissue sarcoma, CCLG

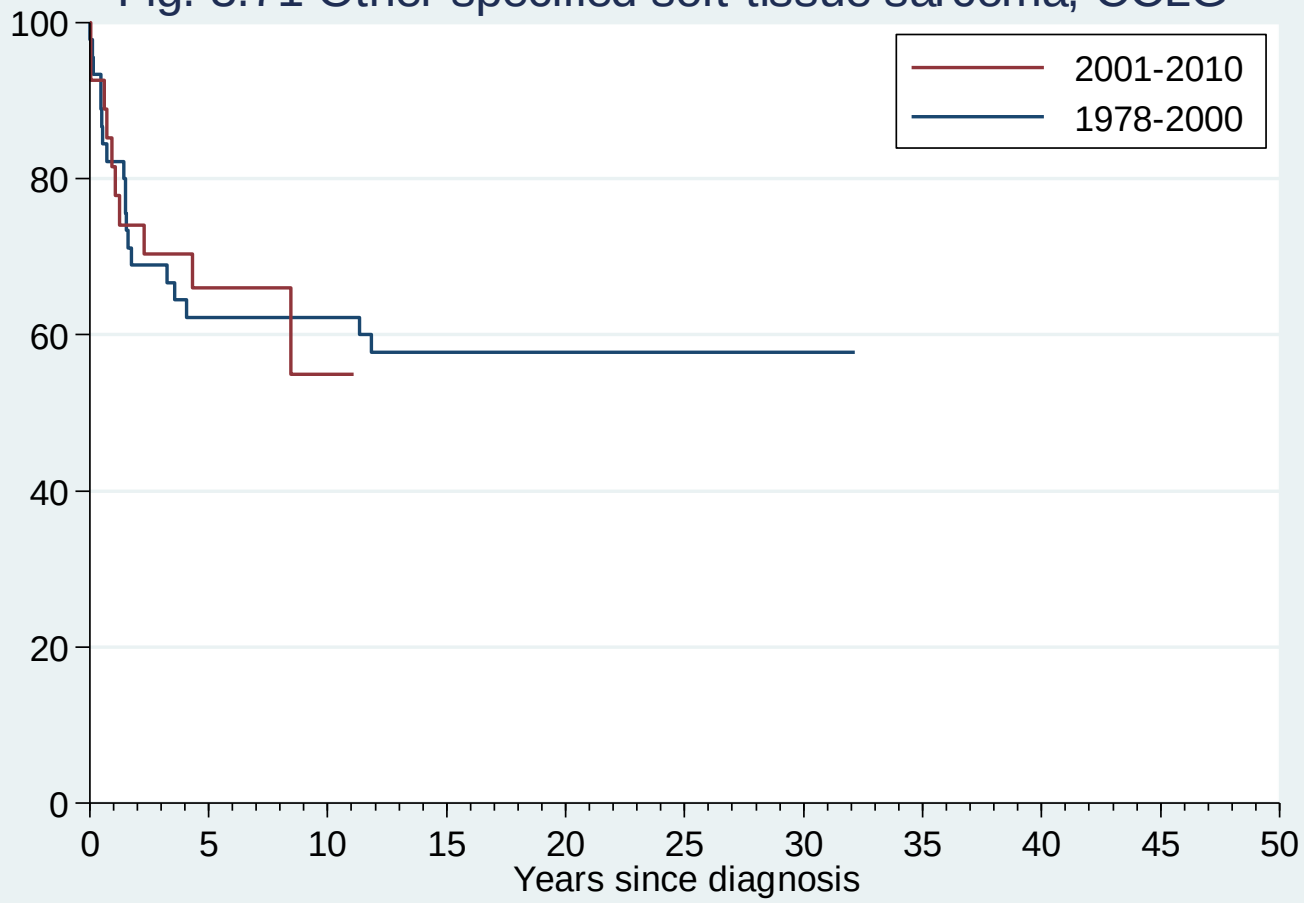


Fig. 3.72 Unspecified soft-tissue sarcoma, CCLG

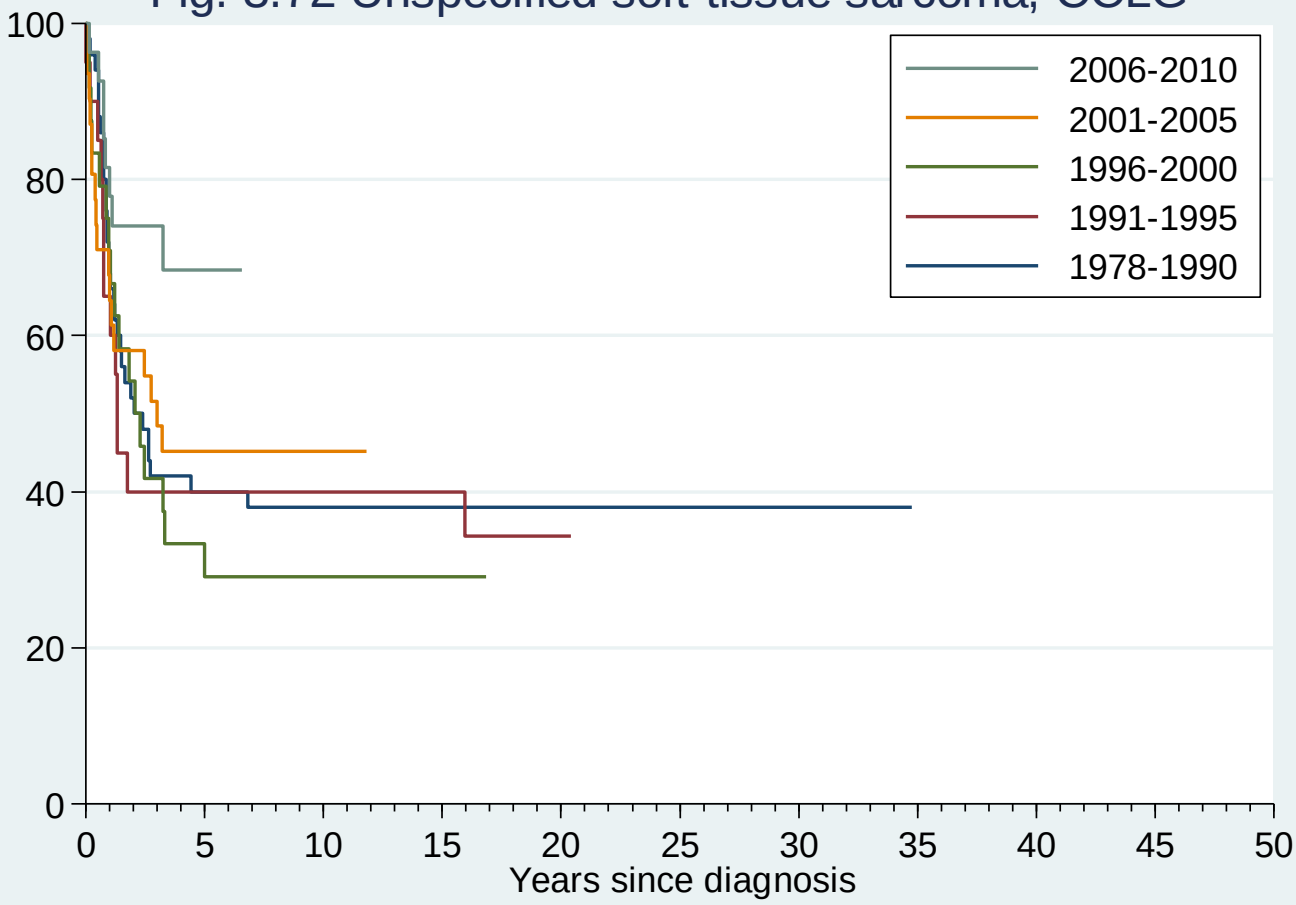


Fig. 3.73 Hepatic sarcoma, CCLG

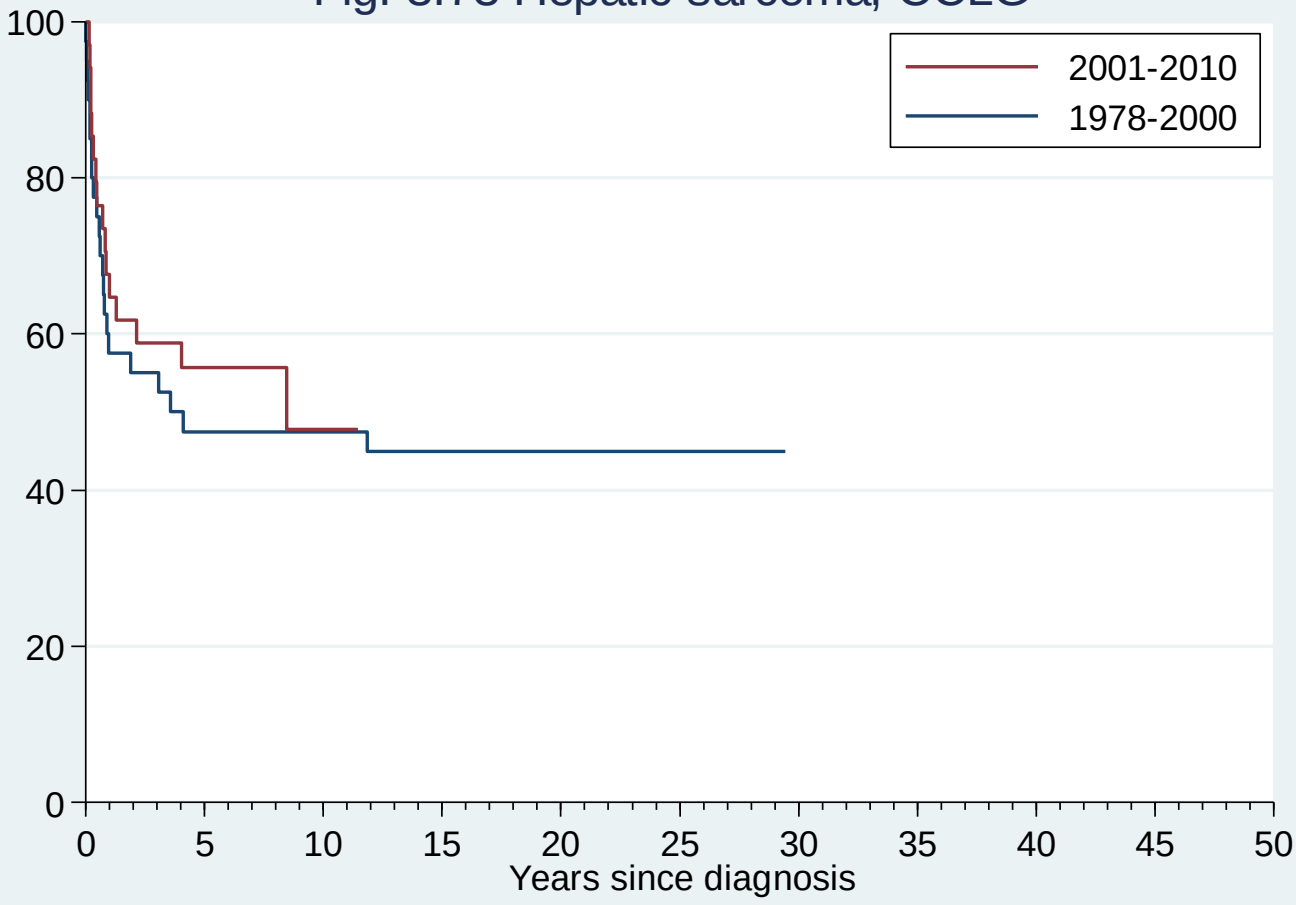


Fig. 3.74 Intracranial and intraspinal germinoma, CCLG

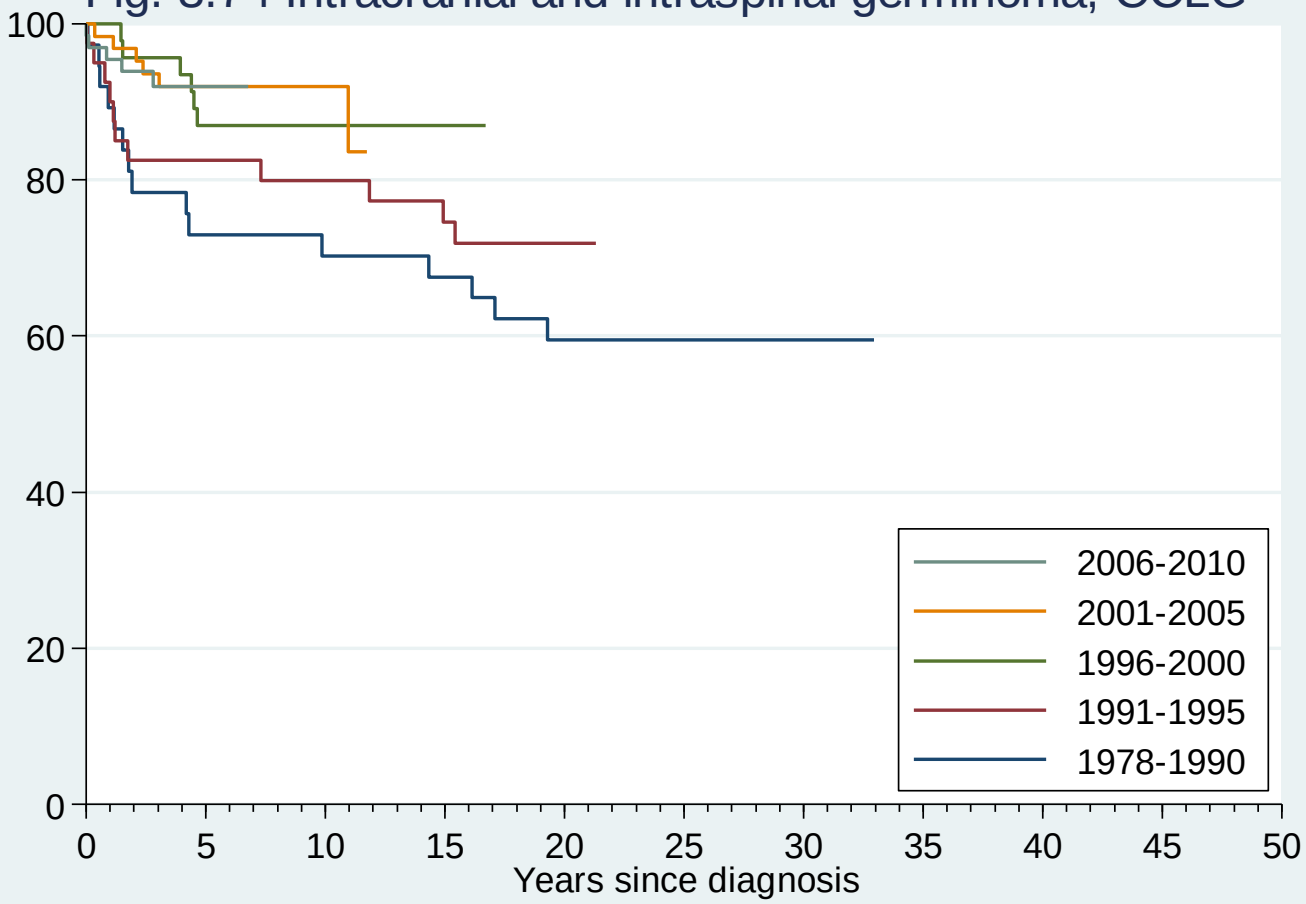


Fig. 3.75 Other intracranial and intraspinal germ-cell, CCLG

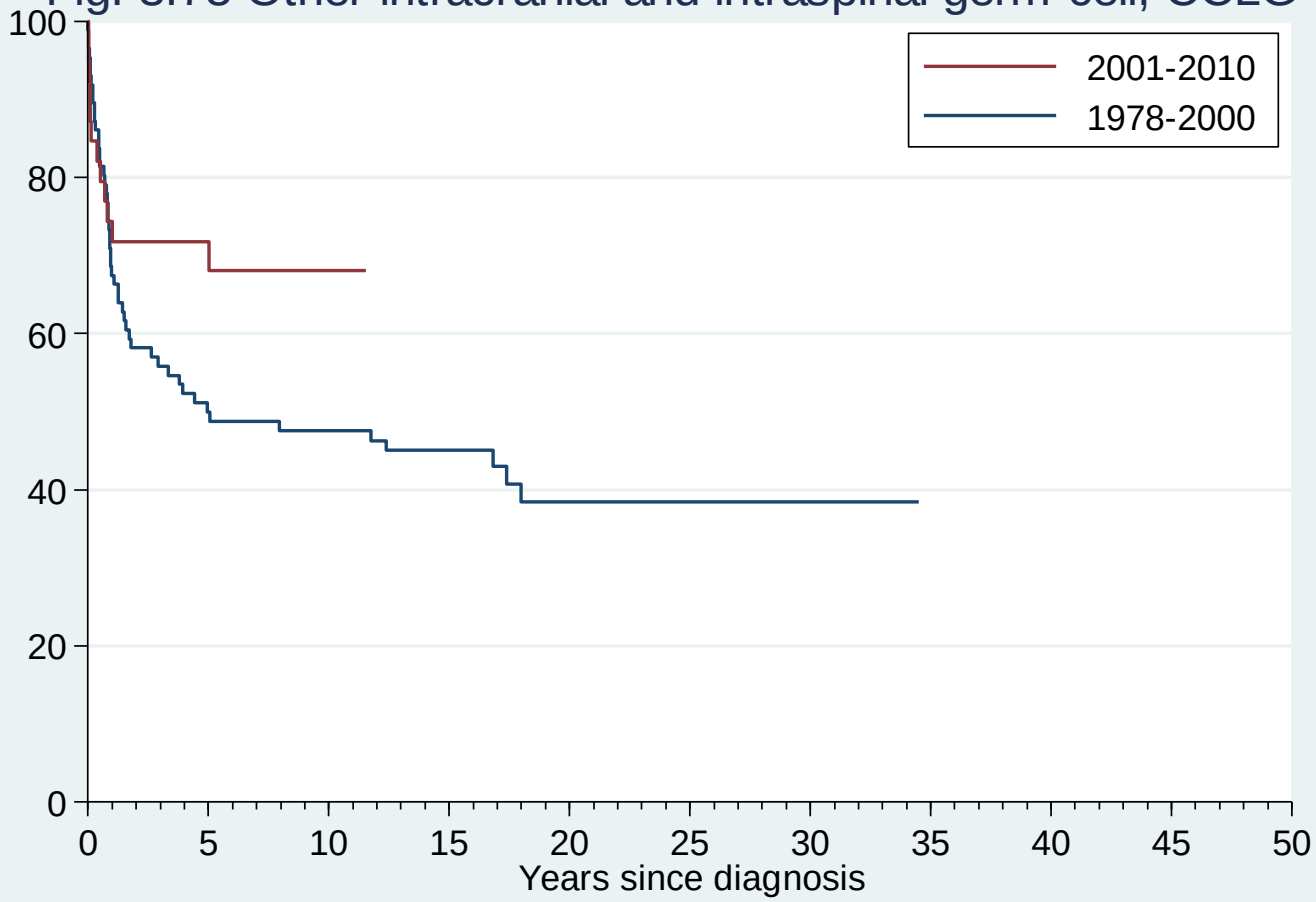


Fig. 3.76 Other malignant extragonadal germ-cell, CCLG

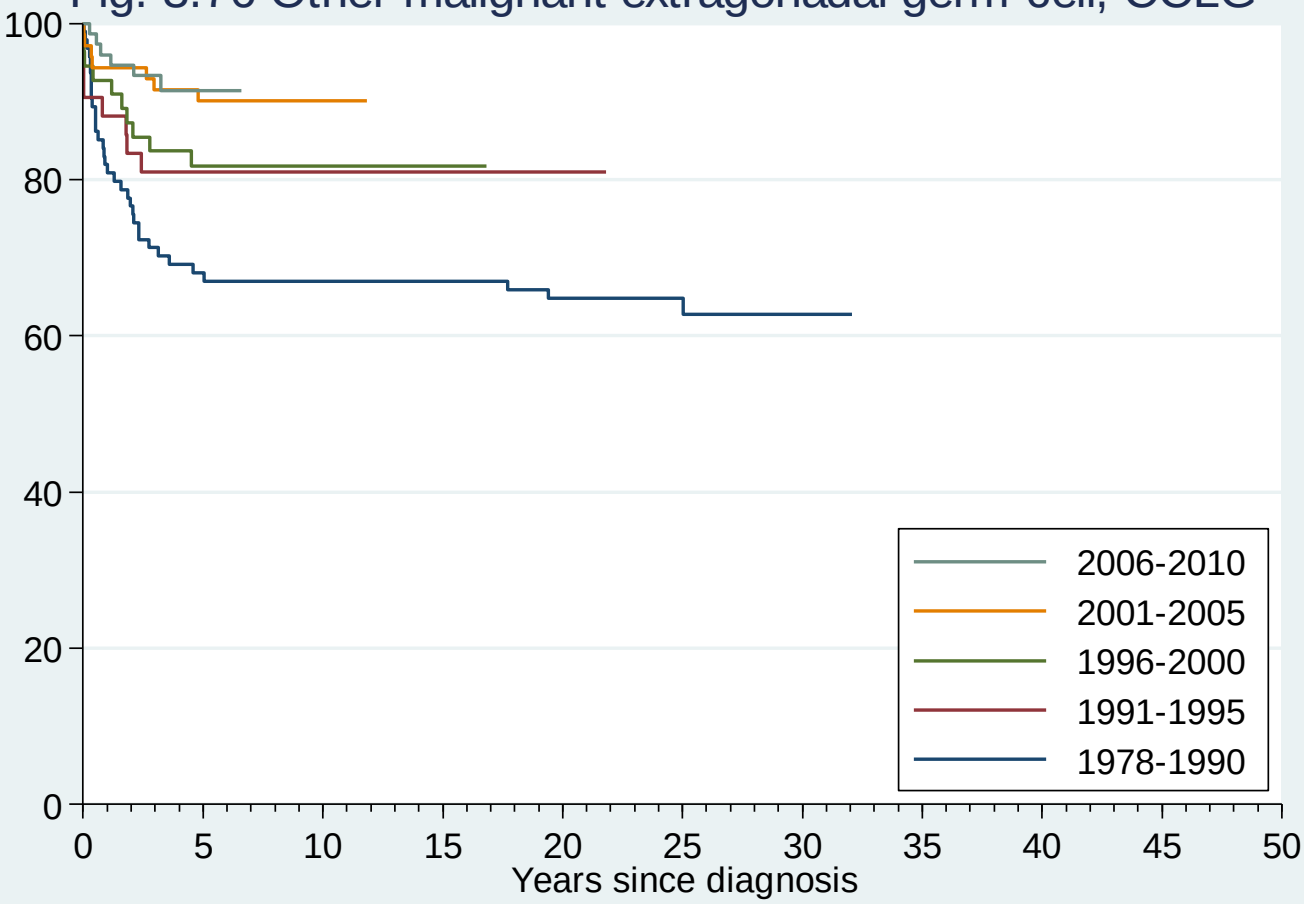


Fig. 3.77 Malignant gonadal germ-cell tumours, CCLG

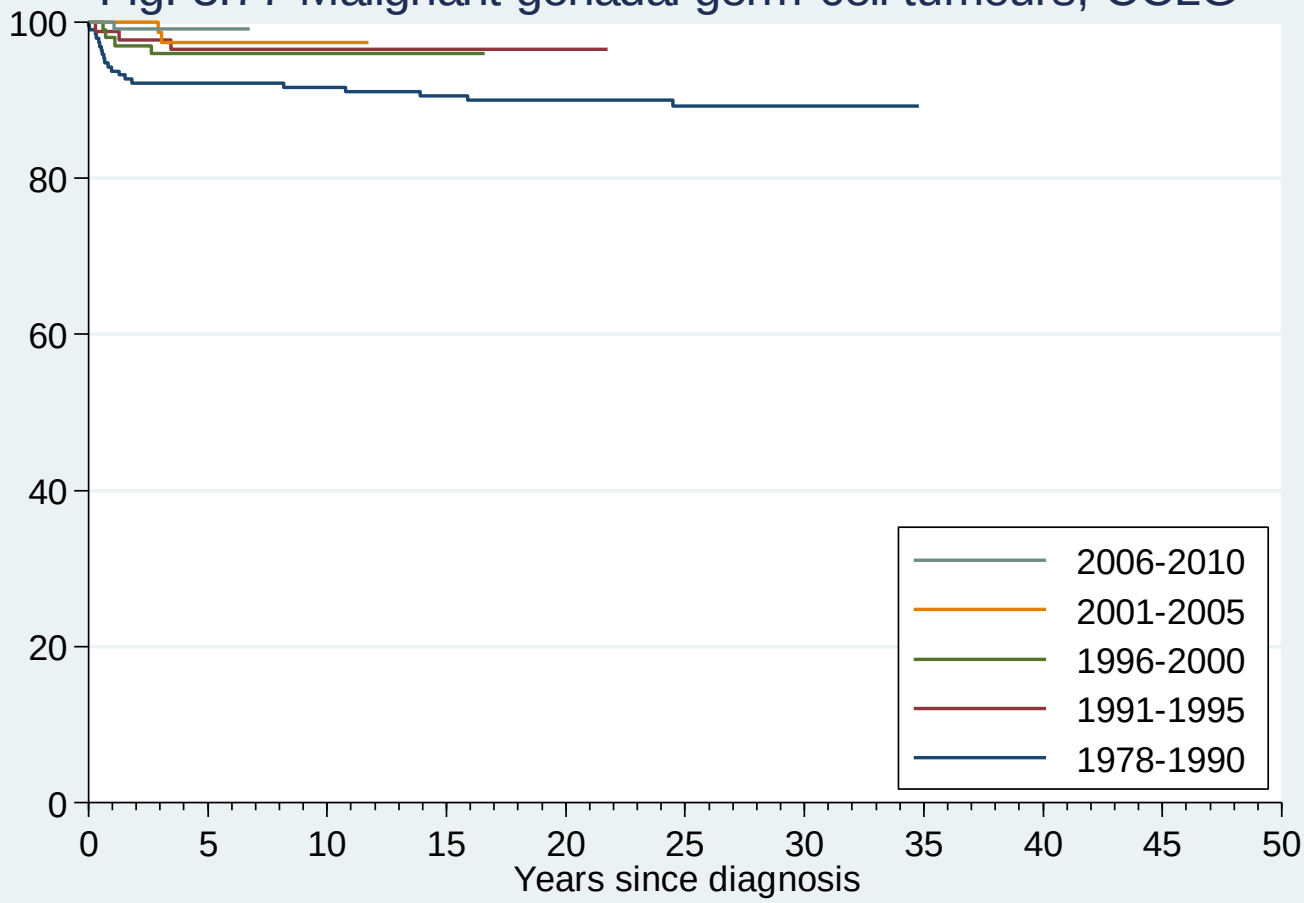


Fig. 3.78 Other malignant gonadal tumours, CCLG

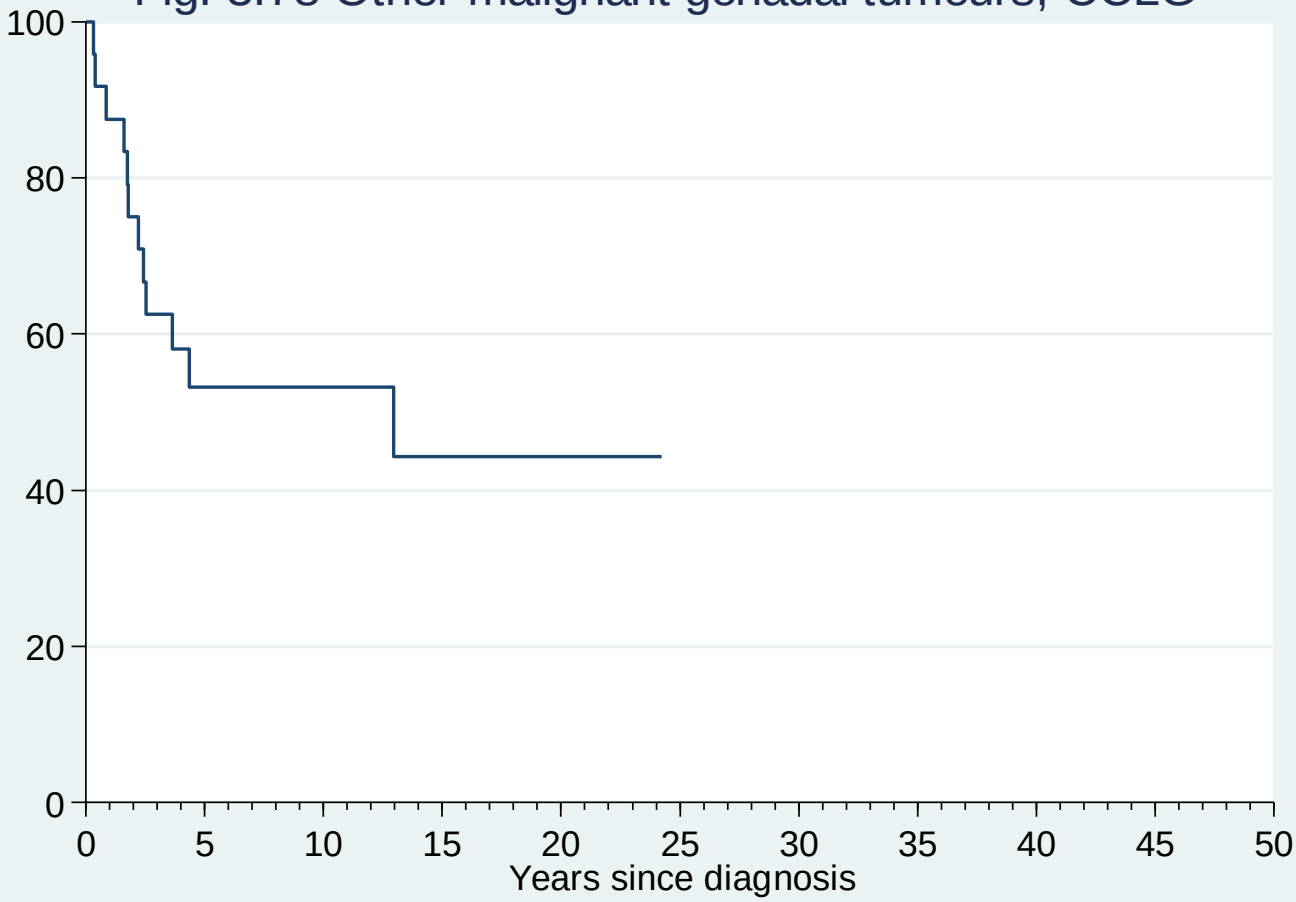


Fig. 3.79 Adrenocortical carcinoma, CCLG

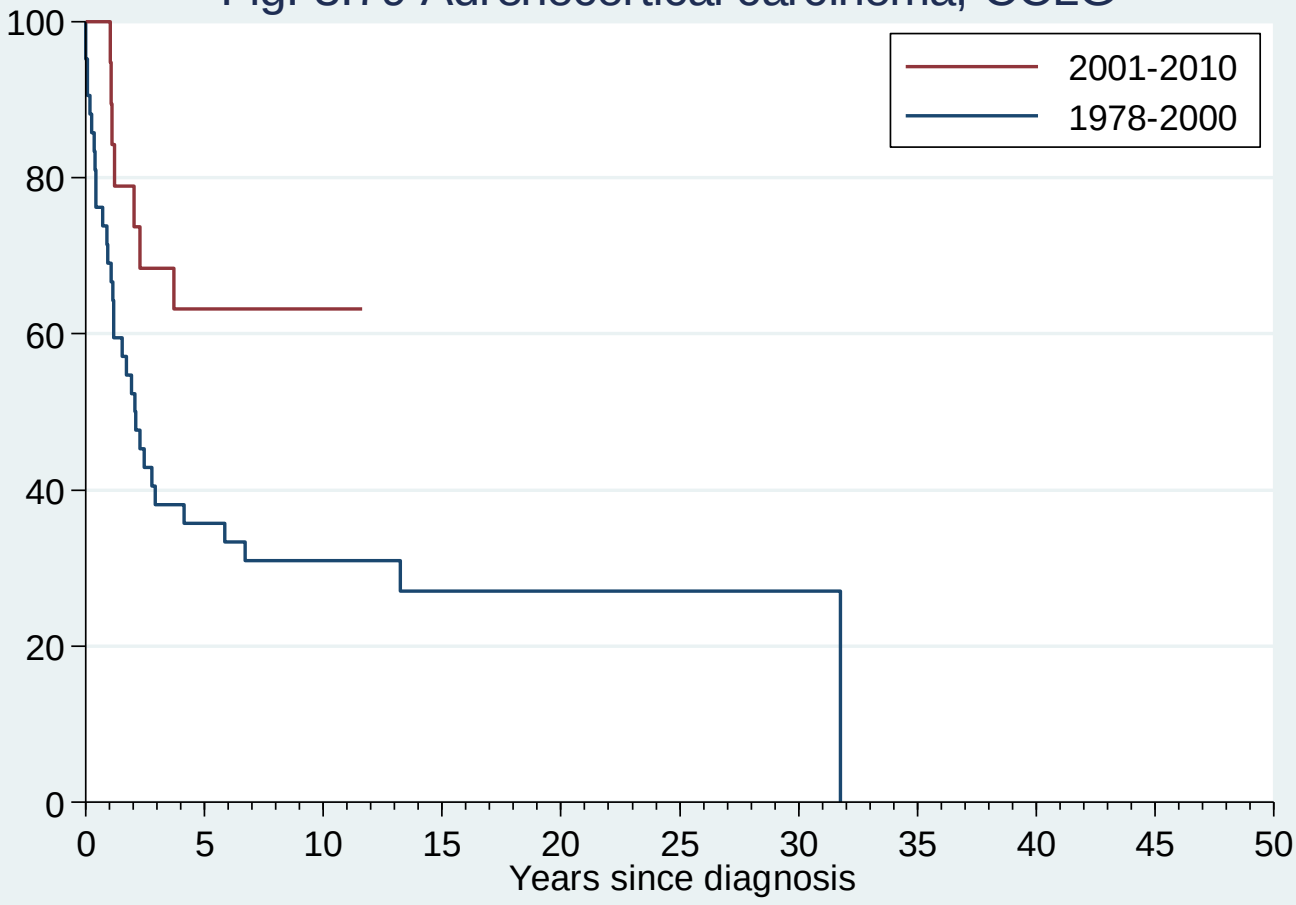


Fig. 3.80 Thyroid carcinoma, non-medullary, CCLG

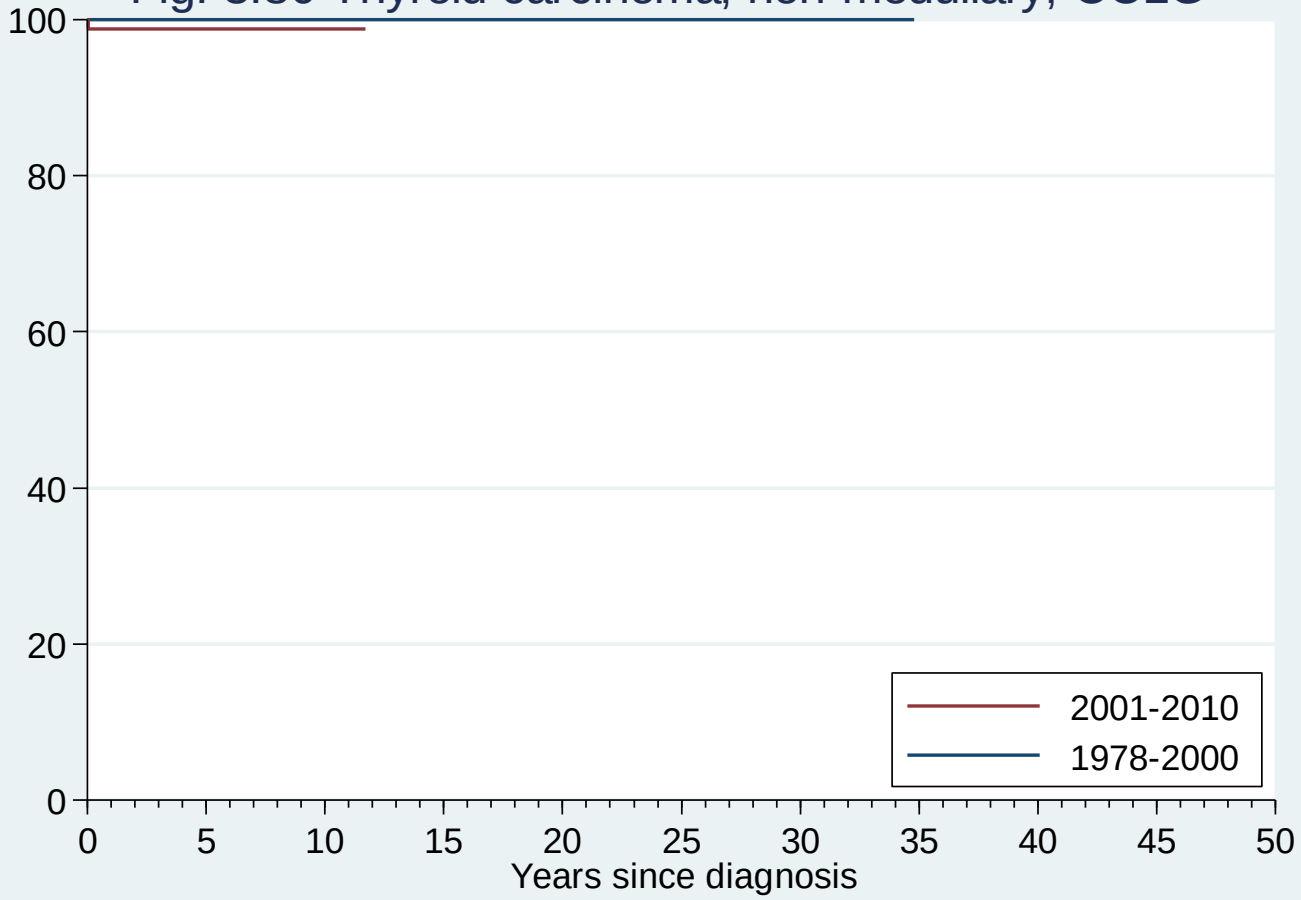


Fig. 3.81 Thyroid carcinoma, medullary, CCLG

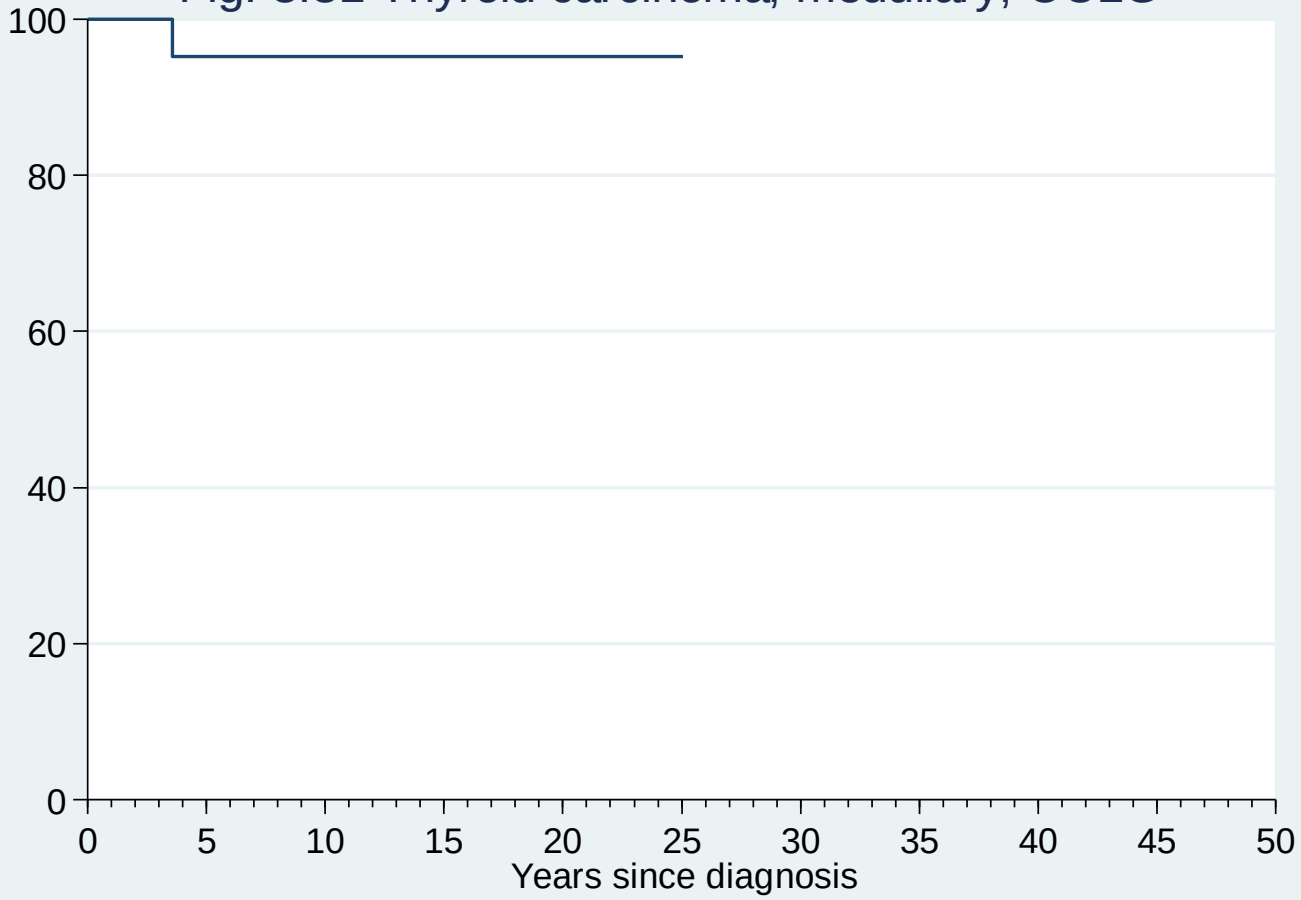


Fig. 3.82 Nasopharyngeal carcinoma, CCLG

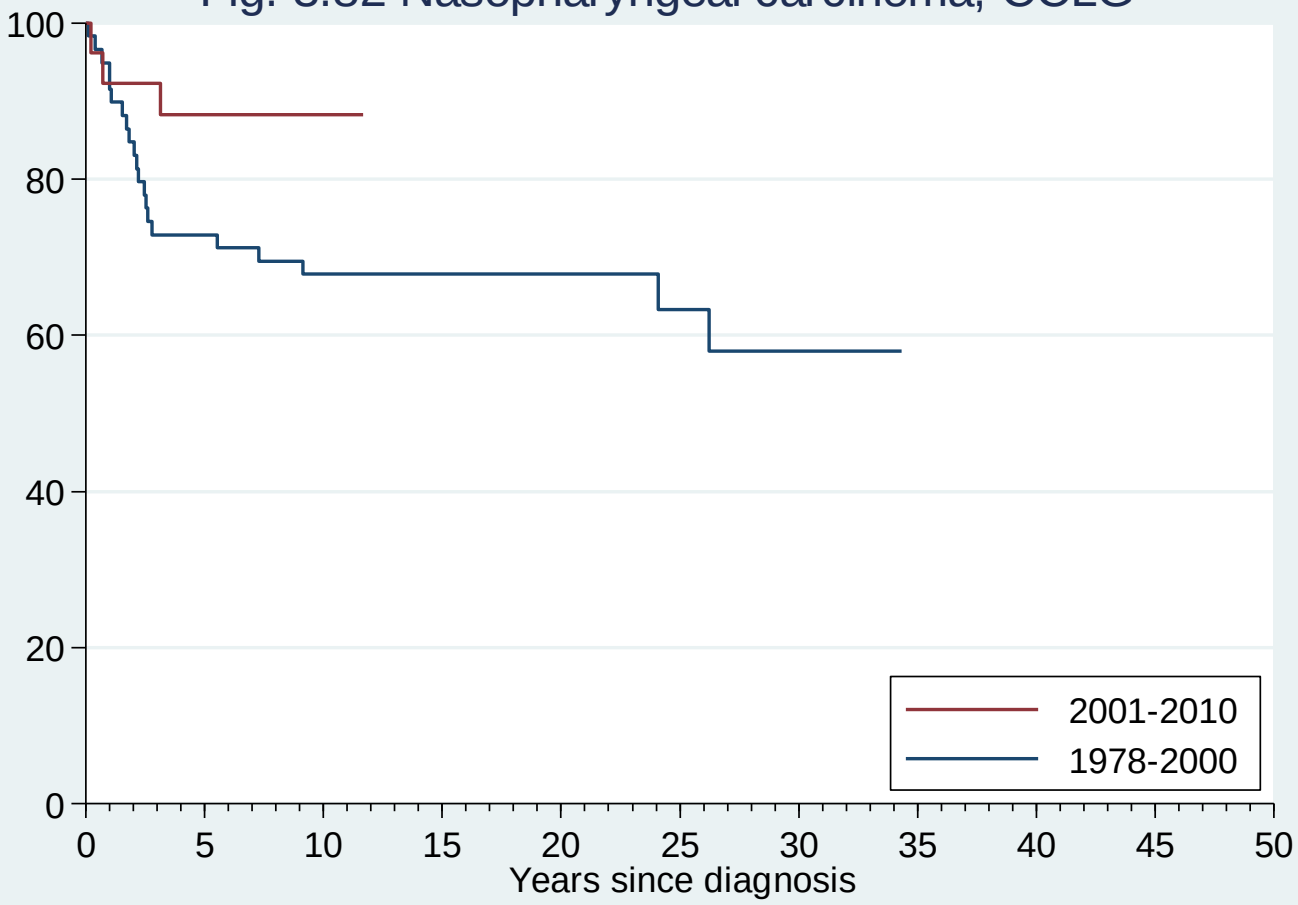


Fig. 3.83 Malignant melanoma, CCLG

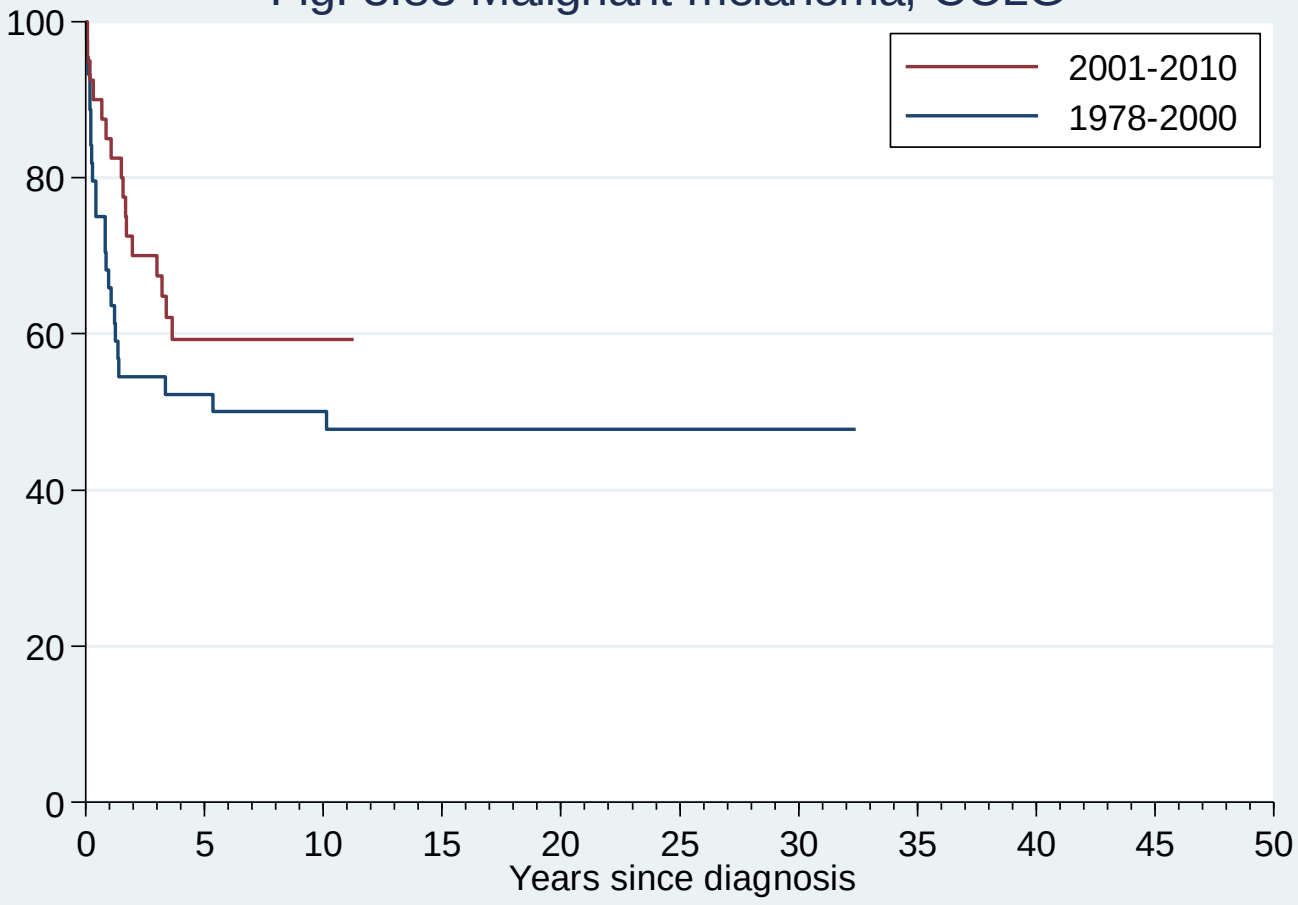


Fig. 3.84 Salivary gland carcinoma, CCLG

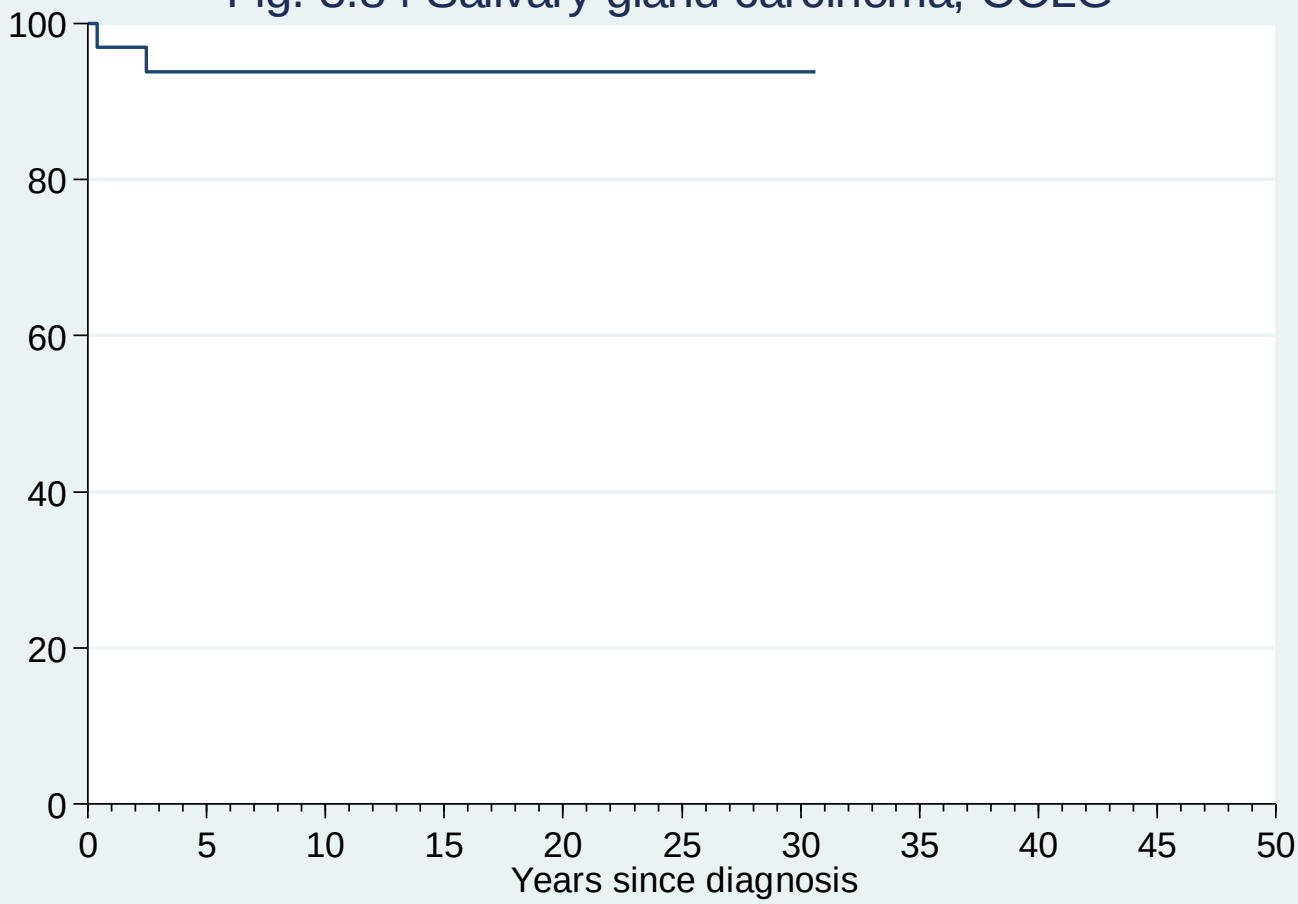


Fig. 3.85 Colorectal carcinoma, CCLG

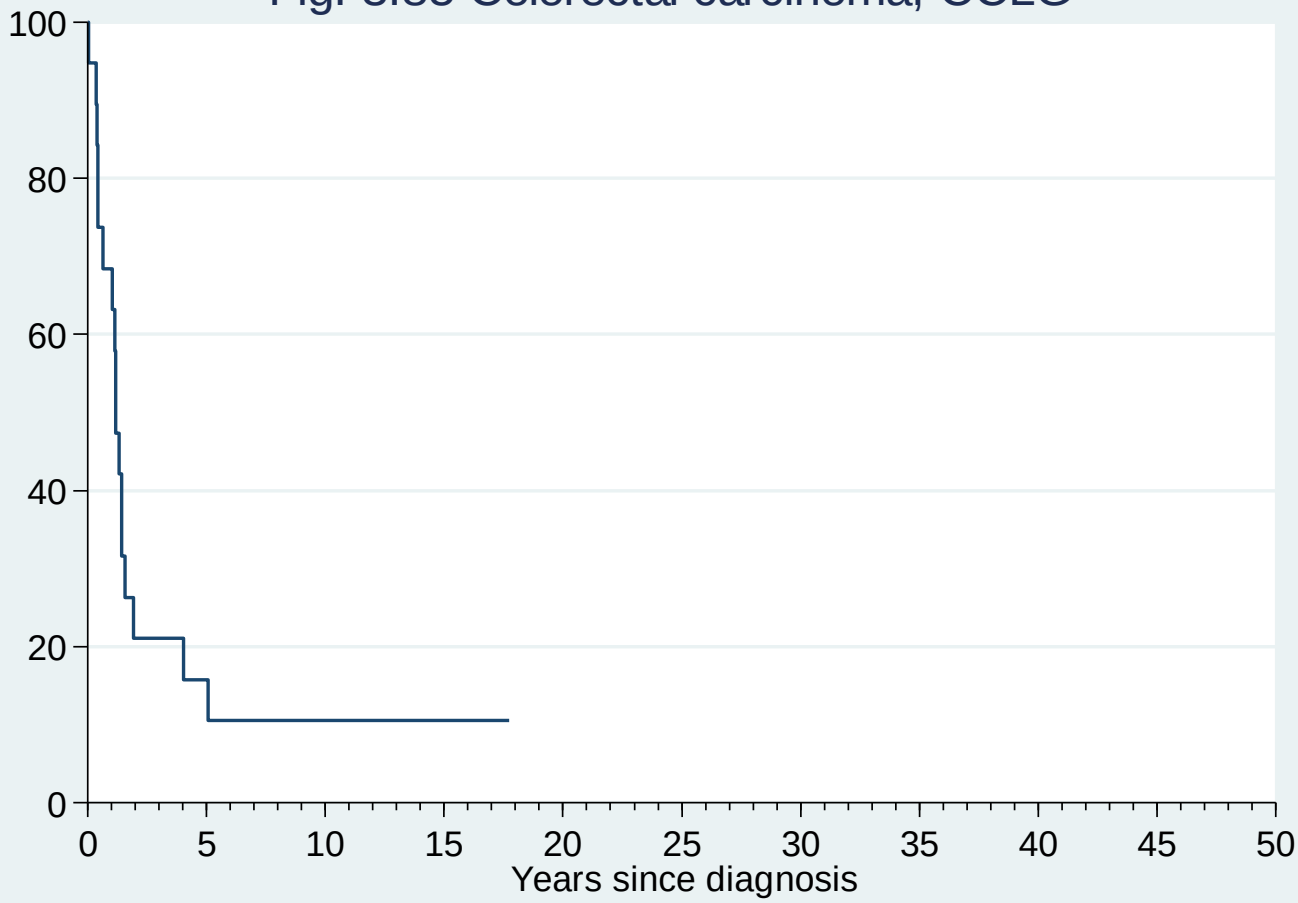


Fig. 3.86 Miscellaneous other carcinoma, CCLG

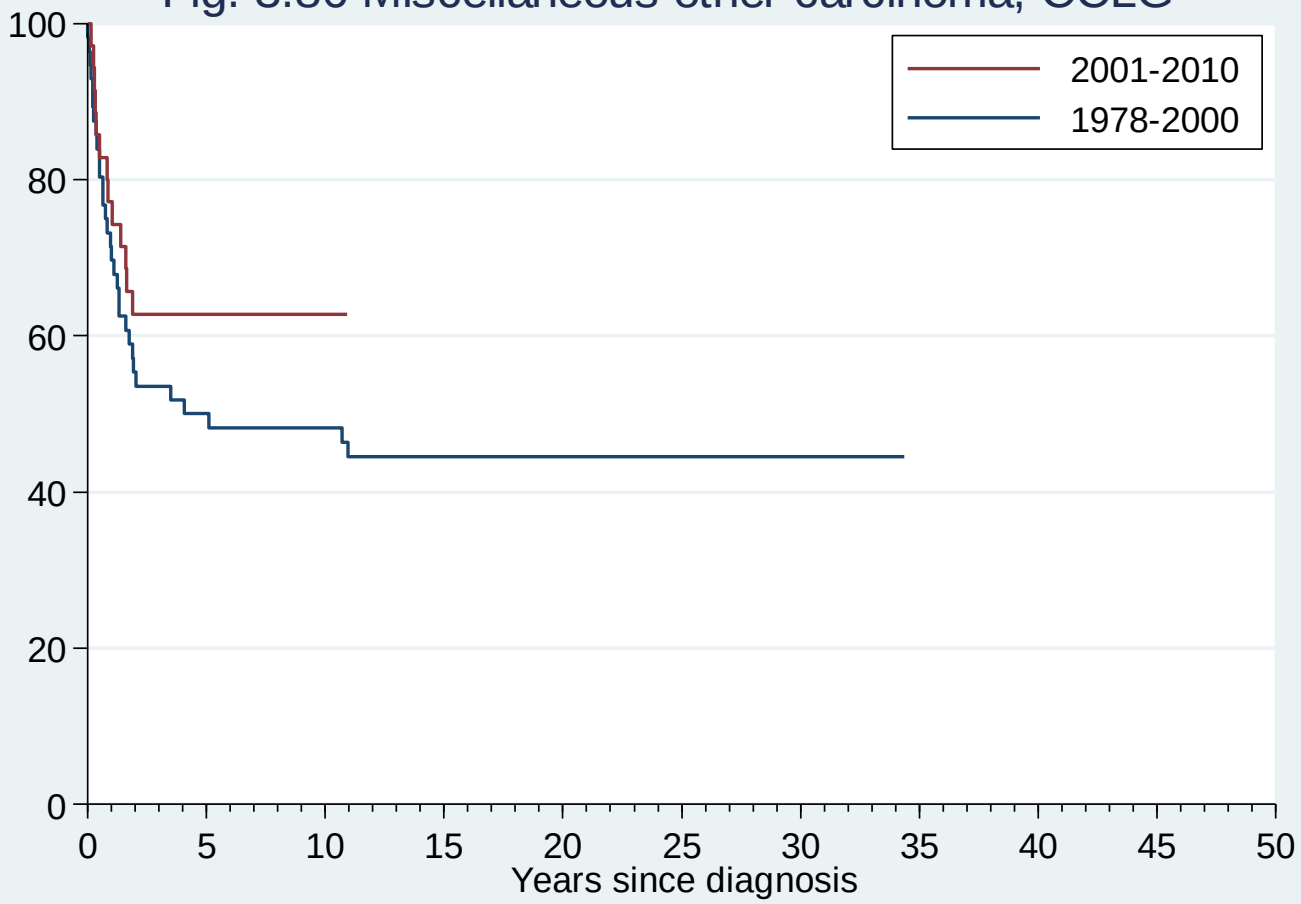


Fig. 3.87 Pleuropulmonary blastoma, CCLG

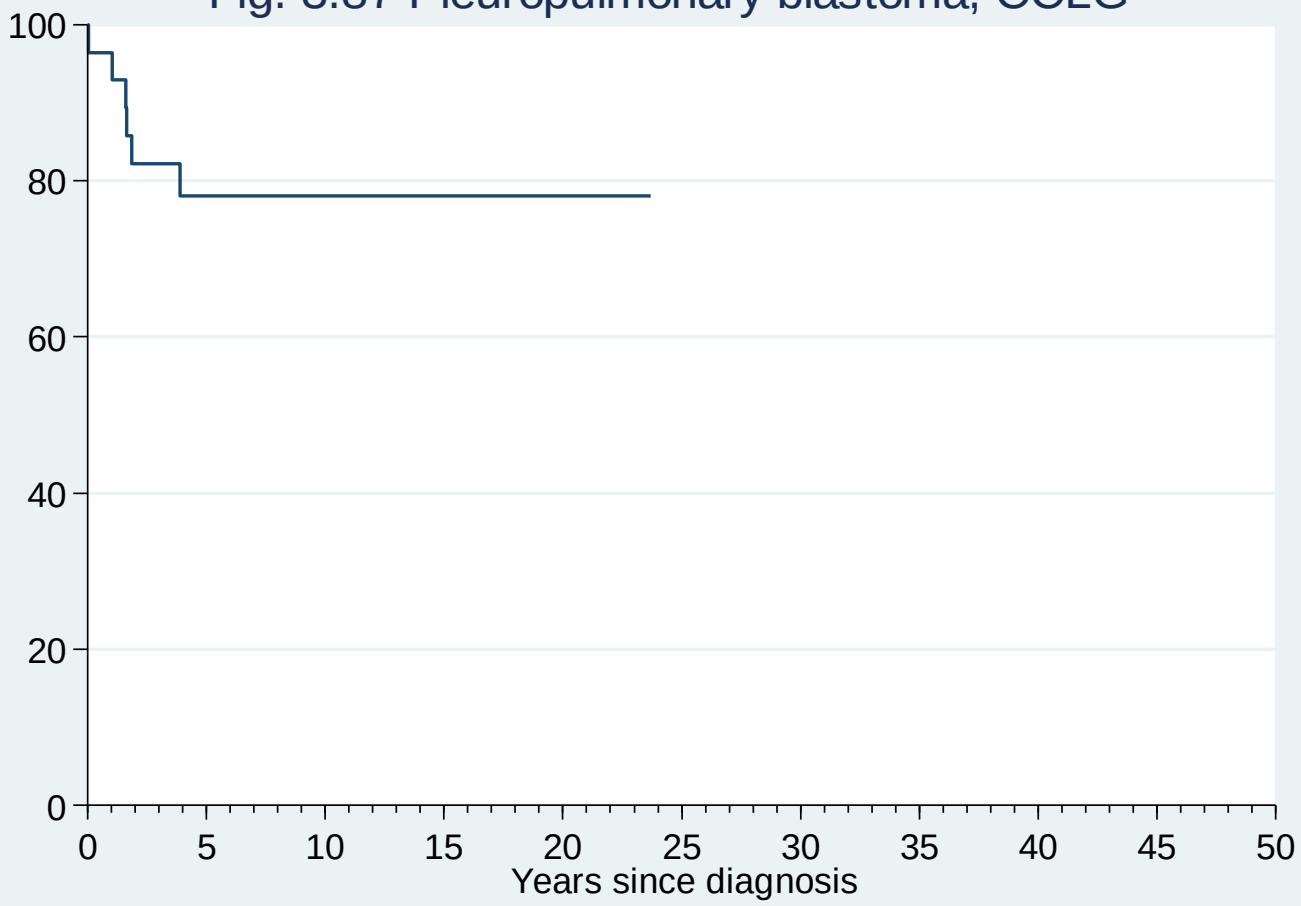


Fig. 3.88 Lymphoproliferative disease, CCLG

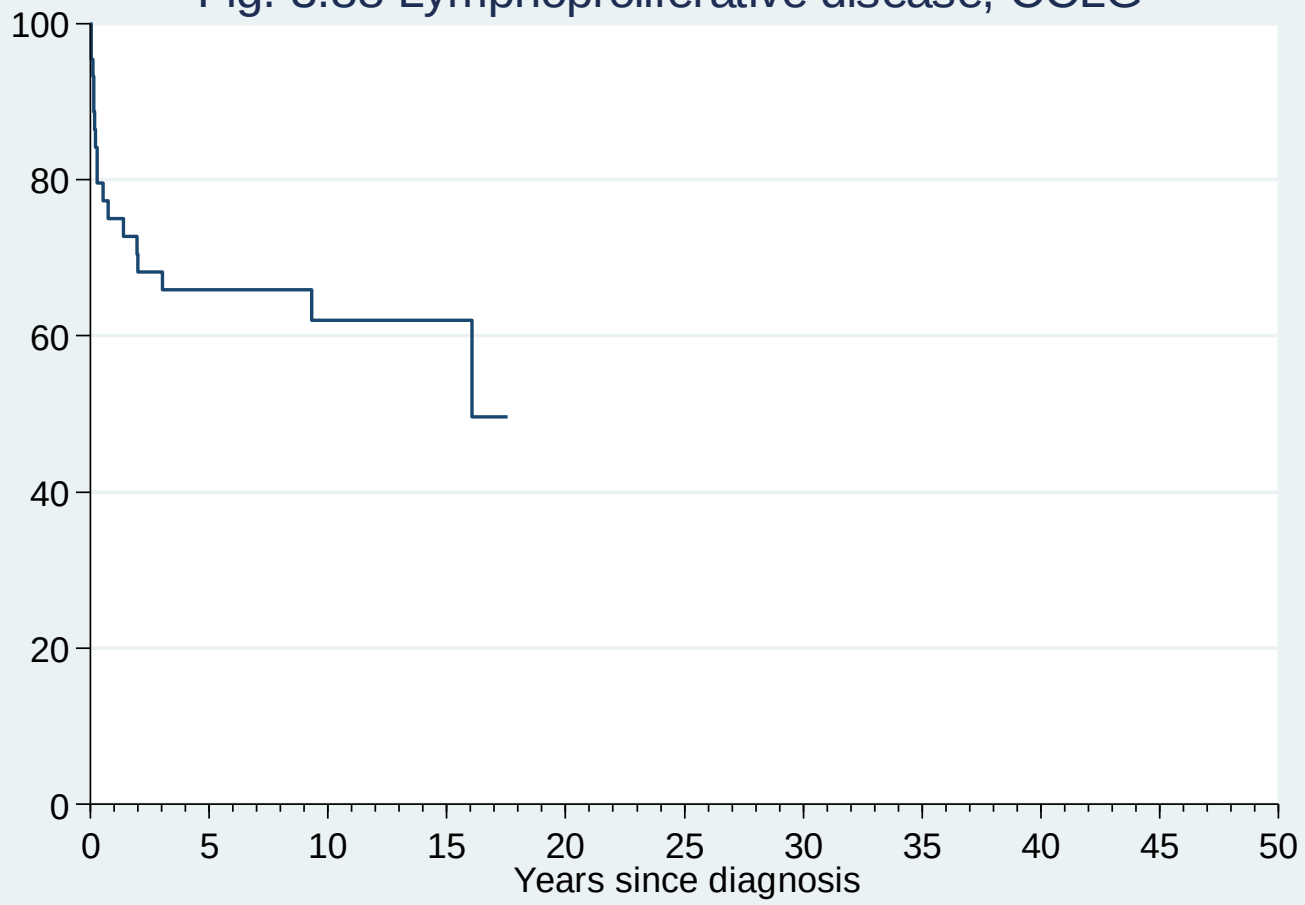


Fig. 3.89 Langerhans cell histiocytosis, single system, CCLG

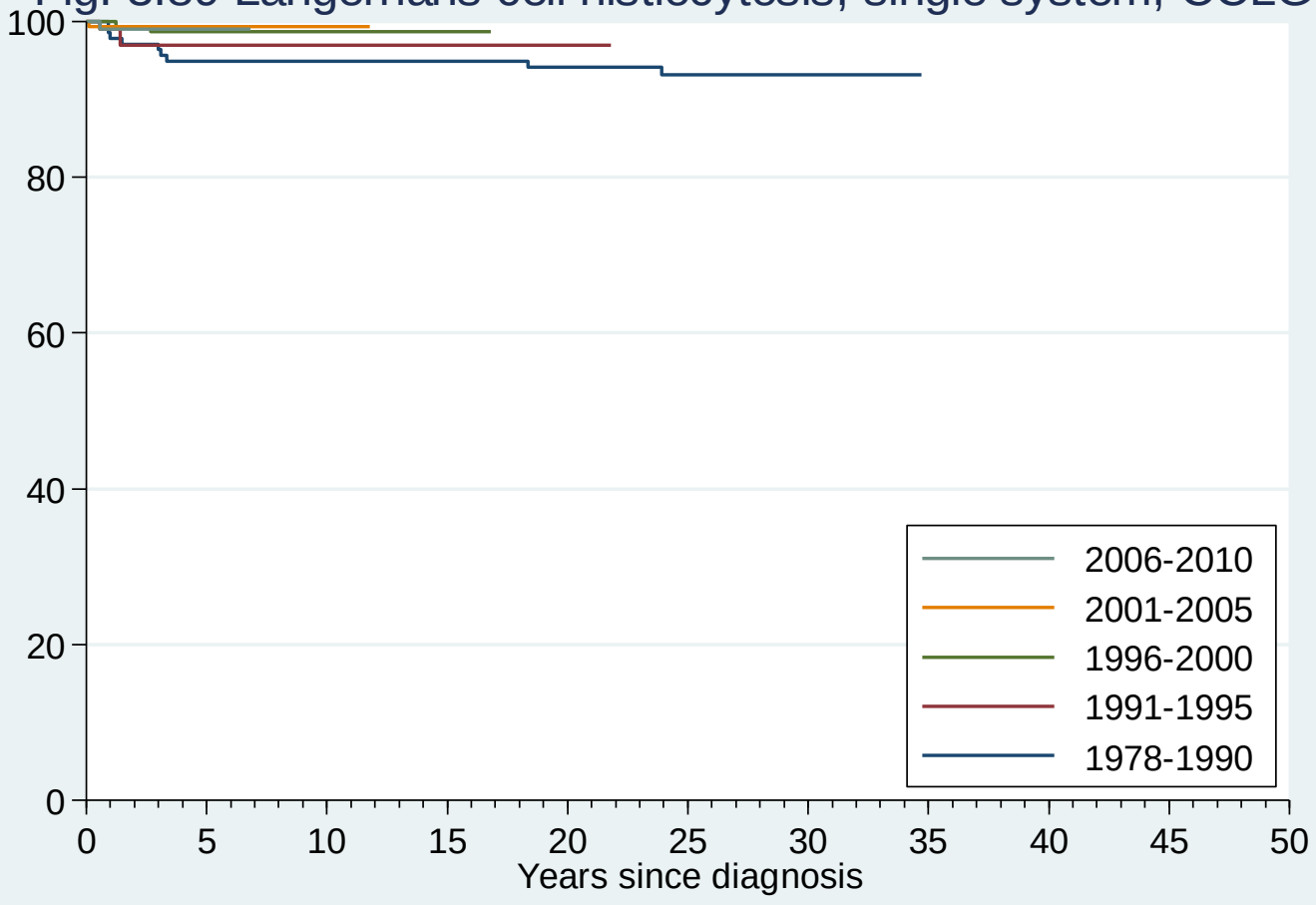


Fig. 3.90 Langerhans cell histiocytosis, multi system, CCLG

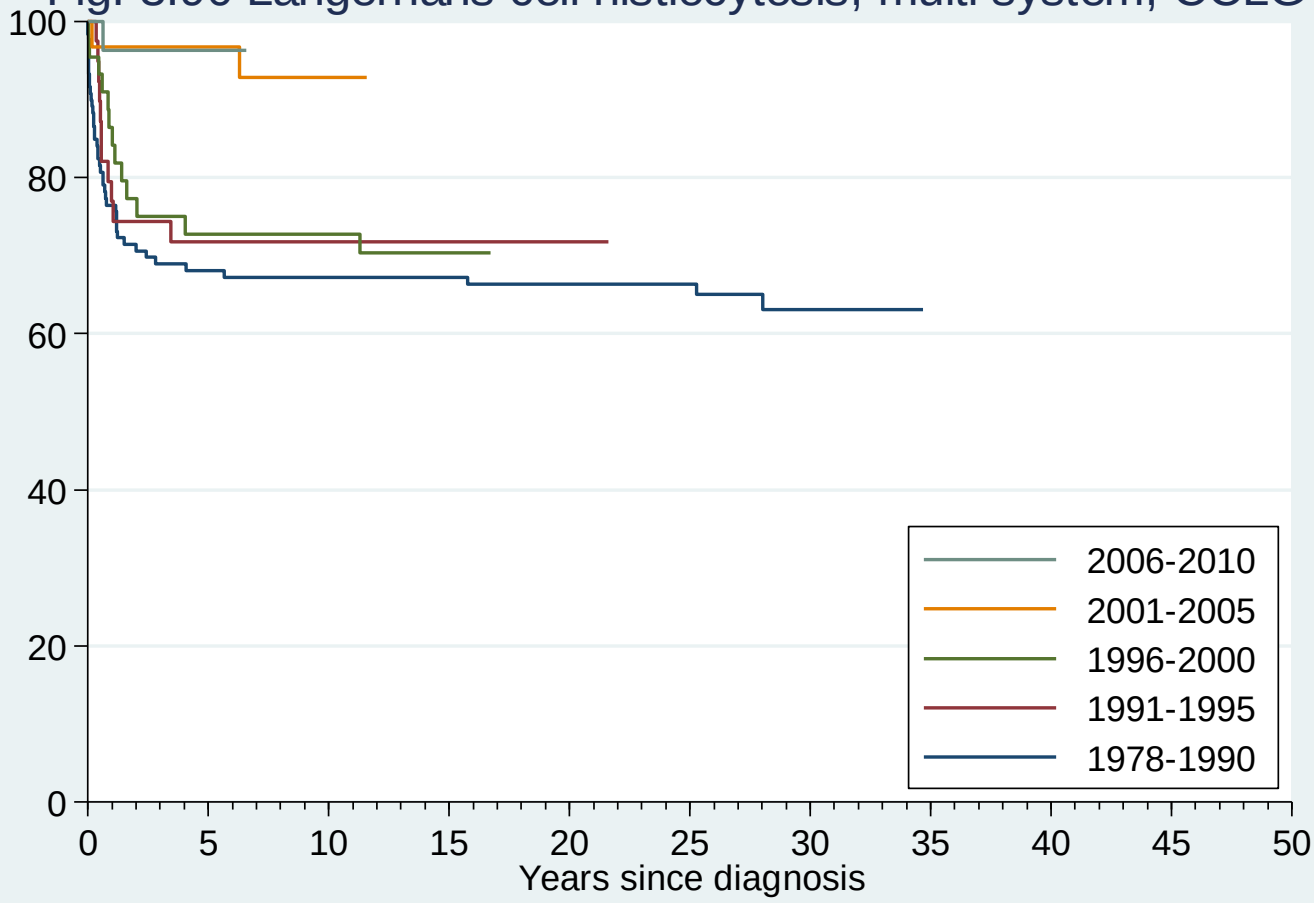


Fig. 3.91 Haemophagocytic lymphohistiocytosis, CCLG

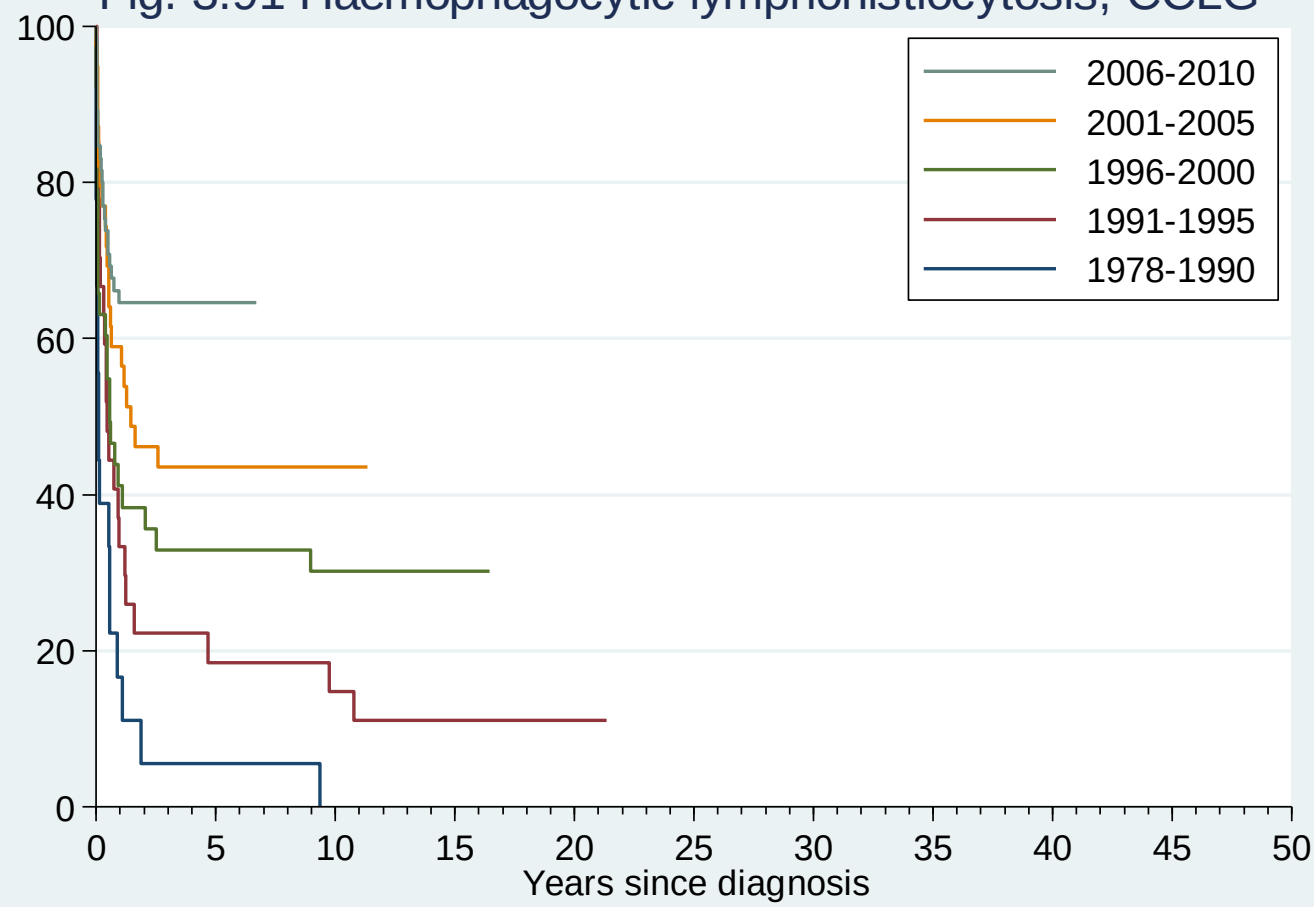


Fig. 3.92 Ganglioneuroma, CCLG

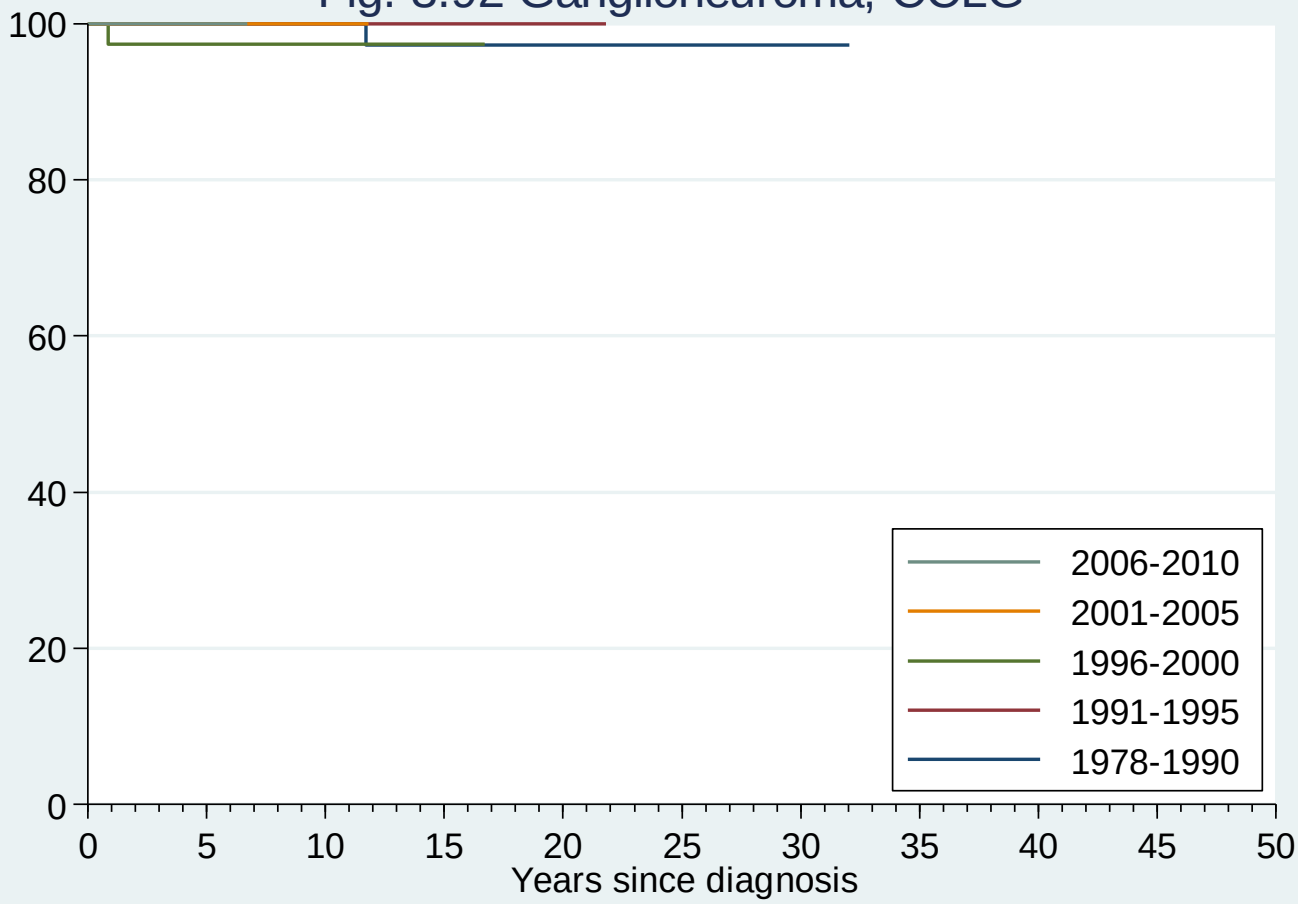


Fig. 3.93 Mesoblastic nephroma, CCLG

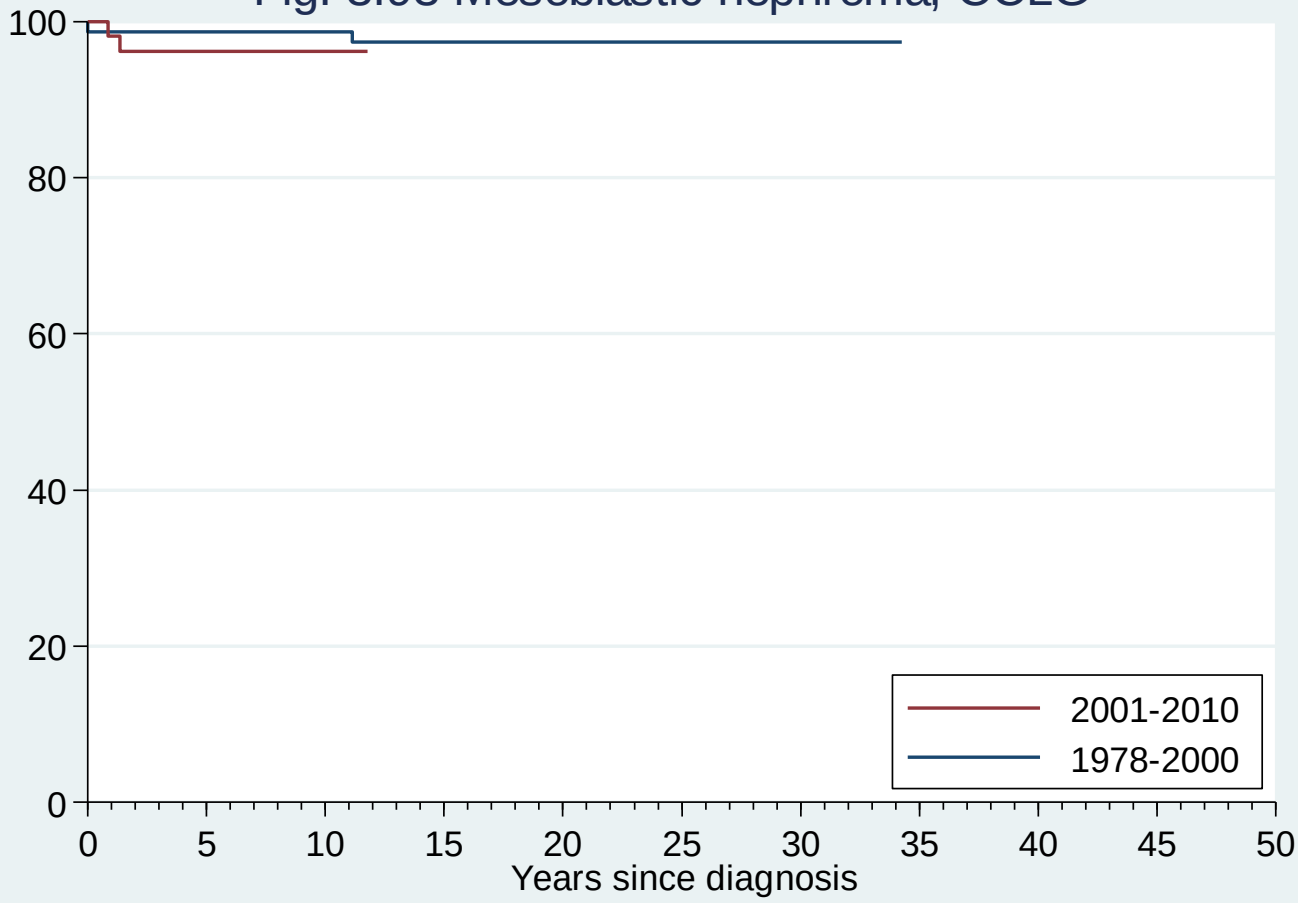


Fig. 3.94 Fibromatosis, CCLG

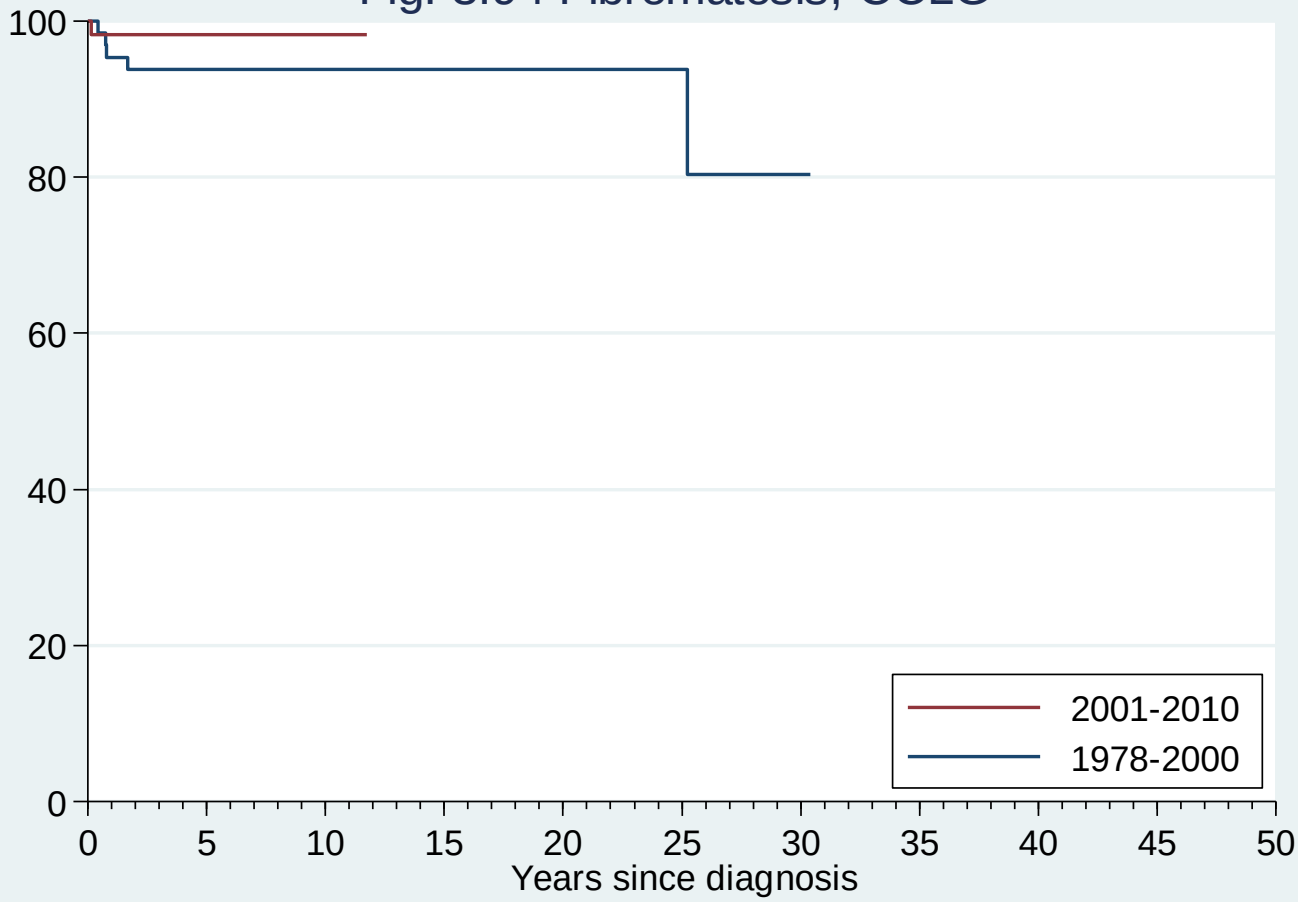


Fig. 3.95 Misc non-malignant soft-tissue tumours, CCLG

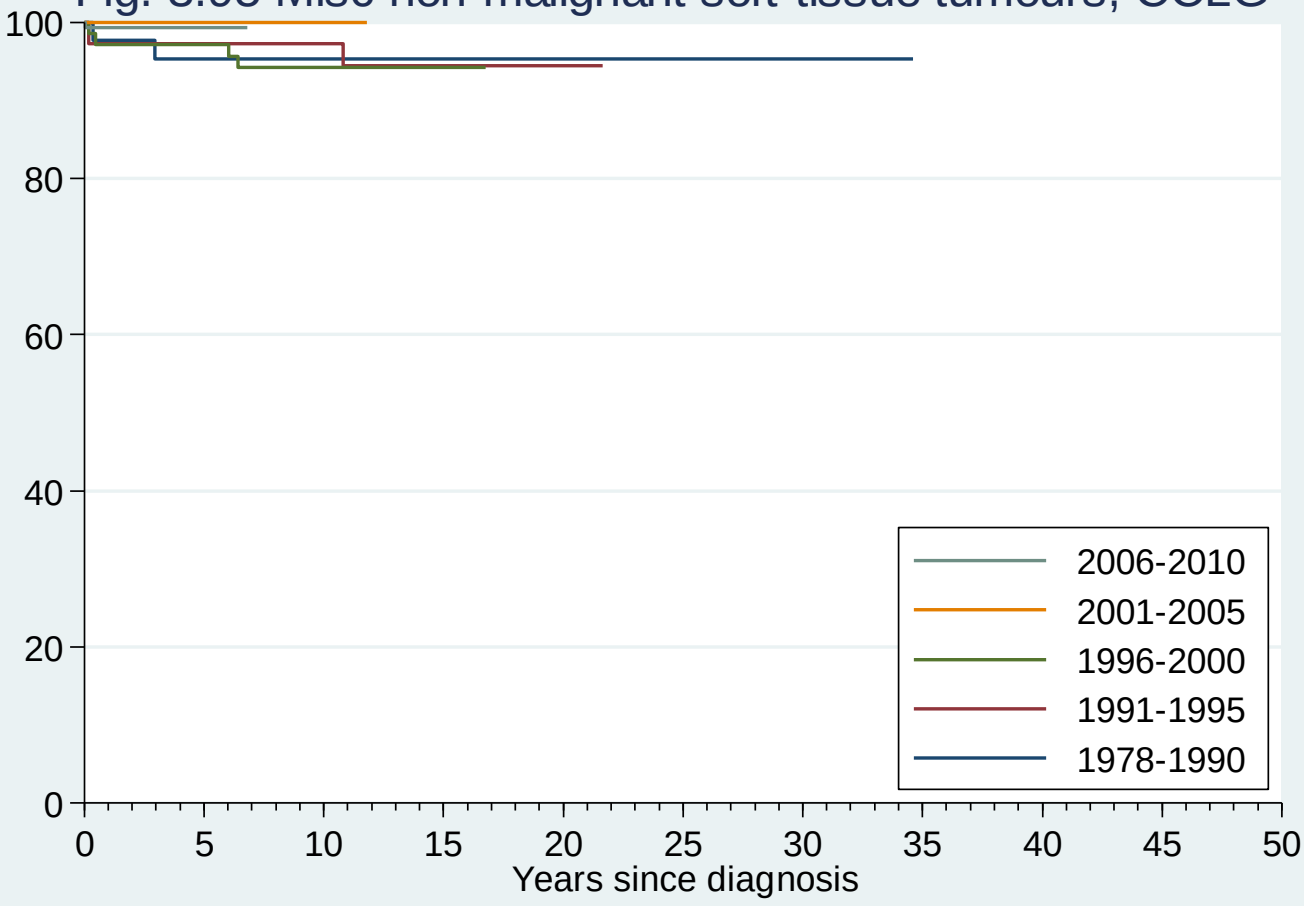


Fig. 3.96 Non-gonadal non-CNS non-malig germ-cell, CCLG

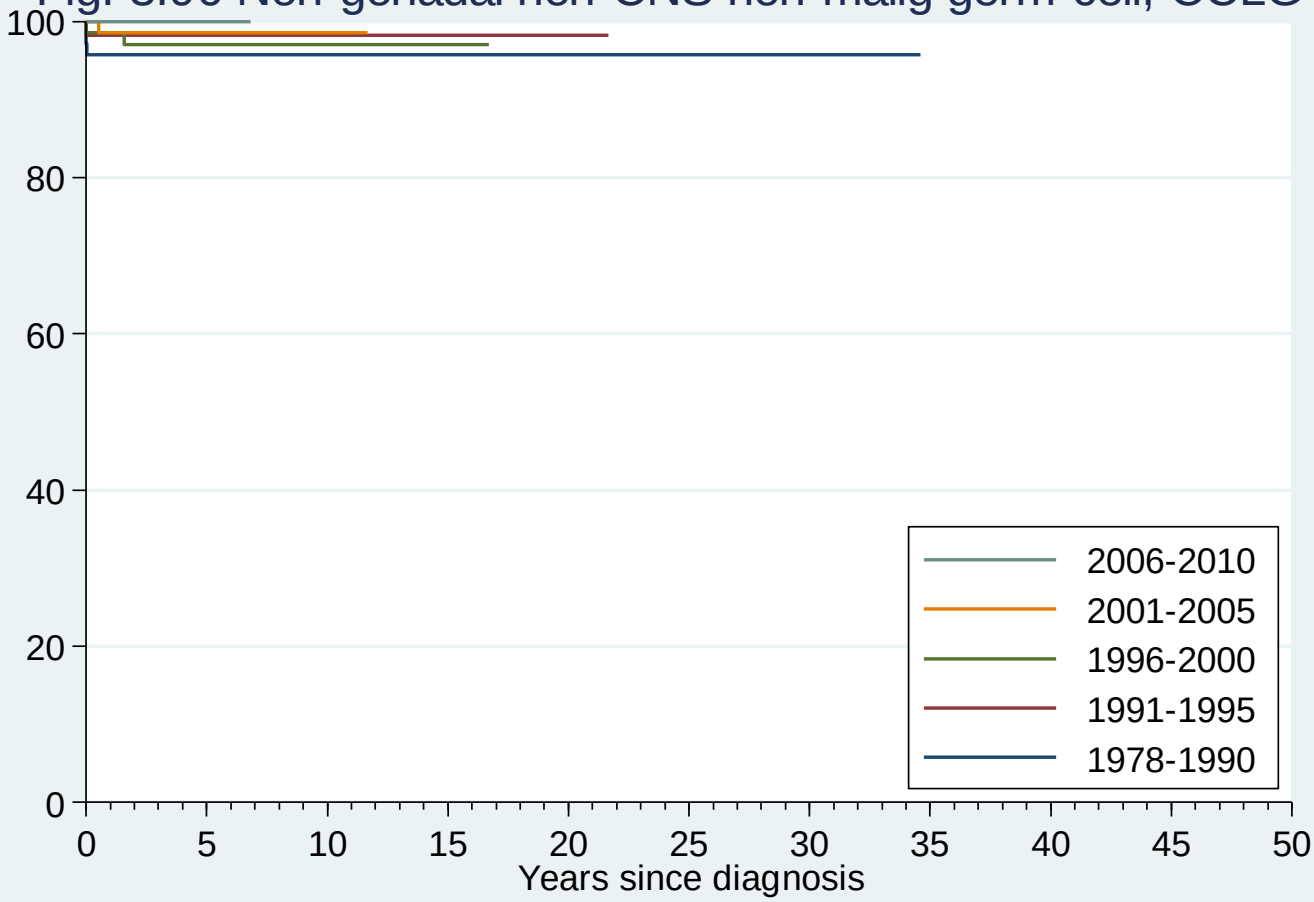


Fig. 3.97 Gonadal non-malignant germ-cell tumours, CCLG

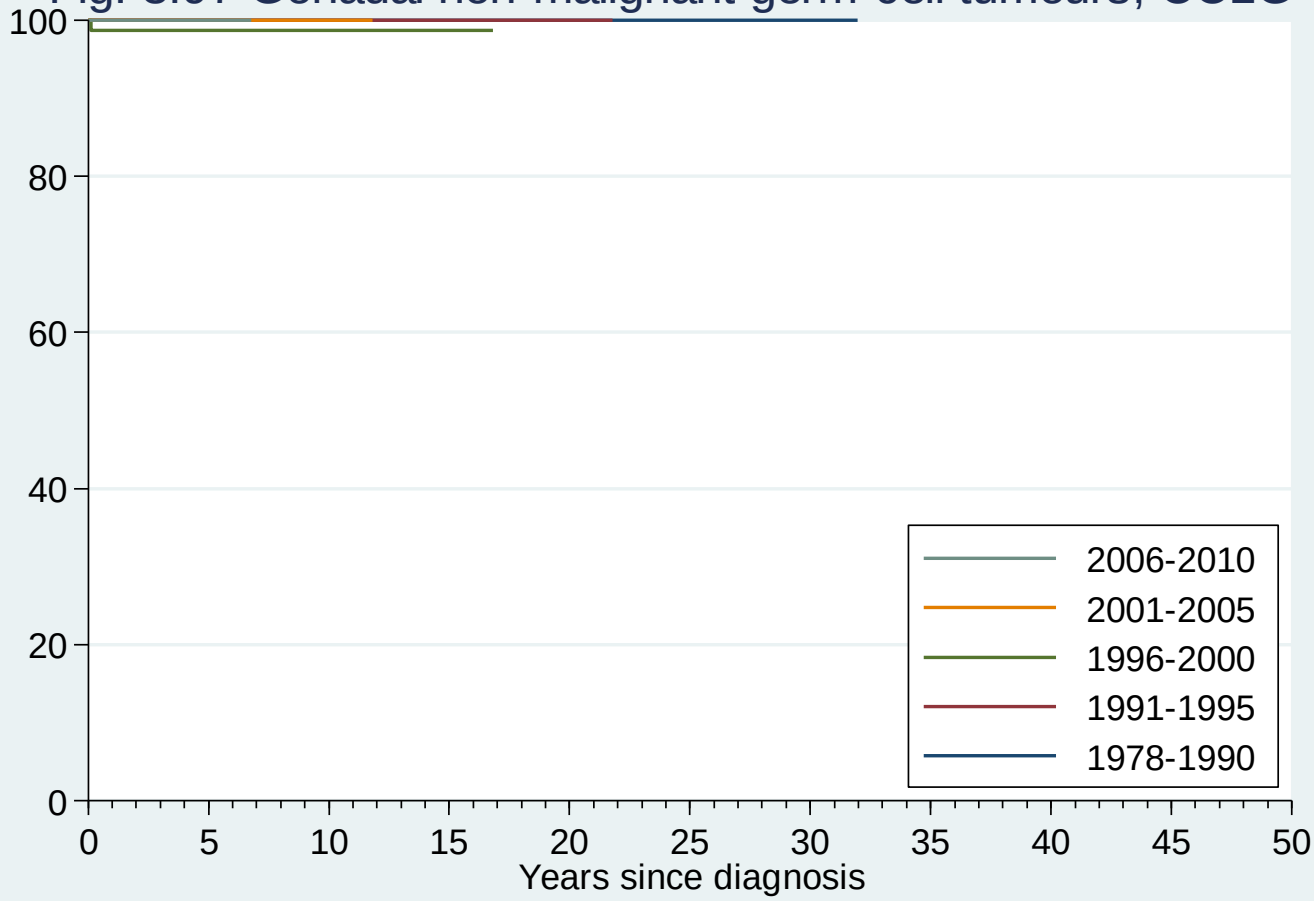


Fig. 3.98 Non-malignant specialised gonadal tumours, CCLG

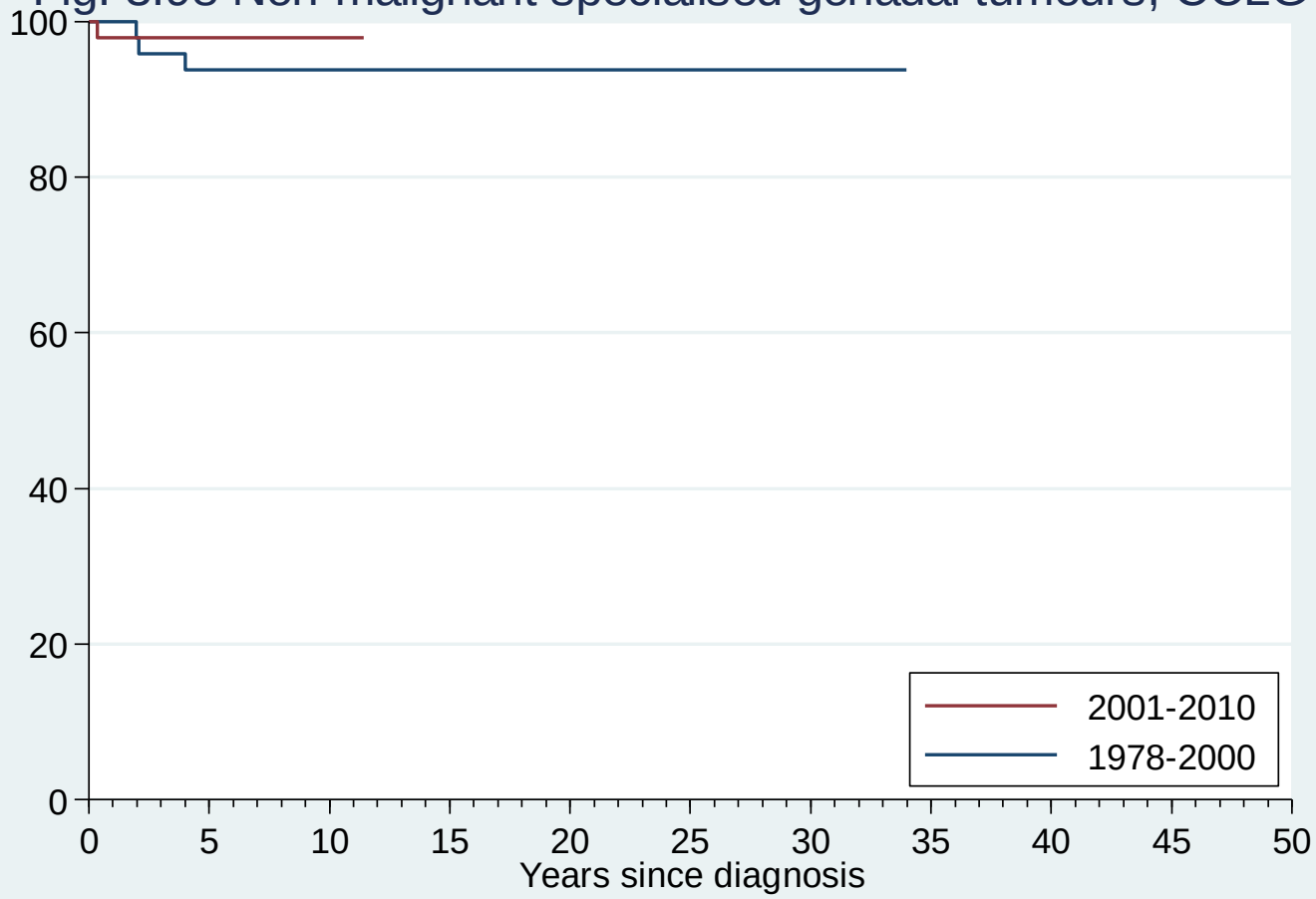


Fig. 3.99 Adrenocortical adenoma, CCLG

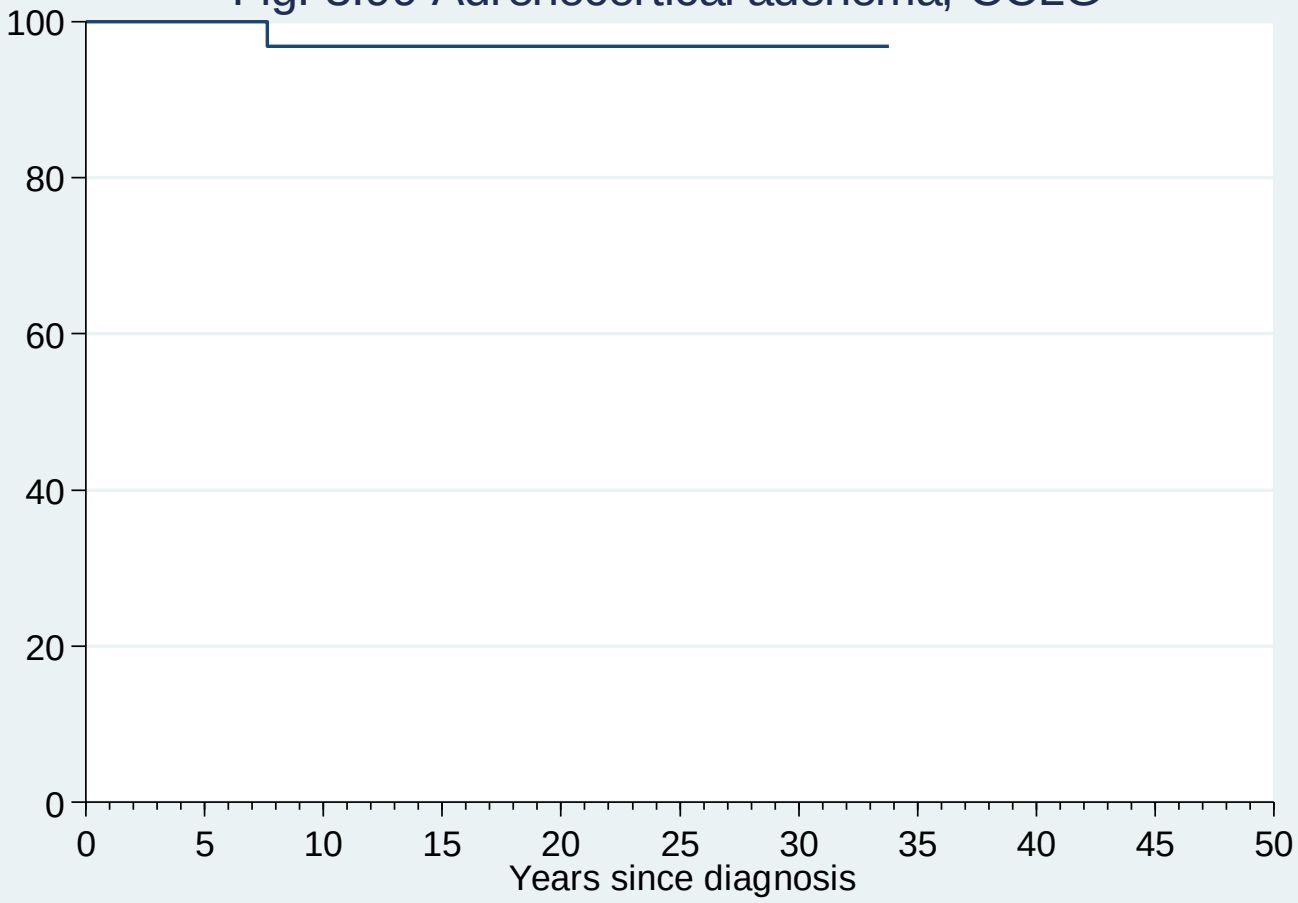


Fig. 3.100 Precursor ALL, non-DS, age < 1 year, CCLG

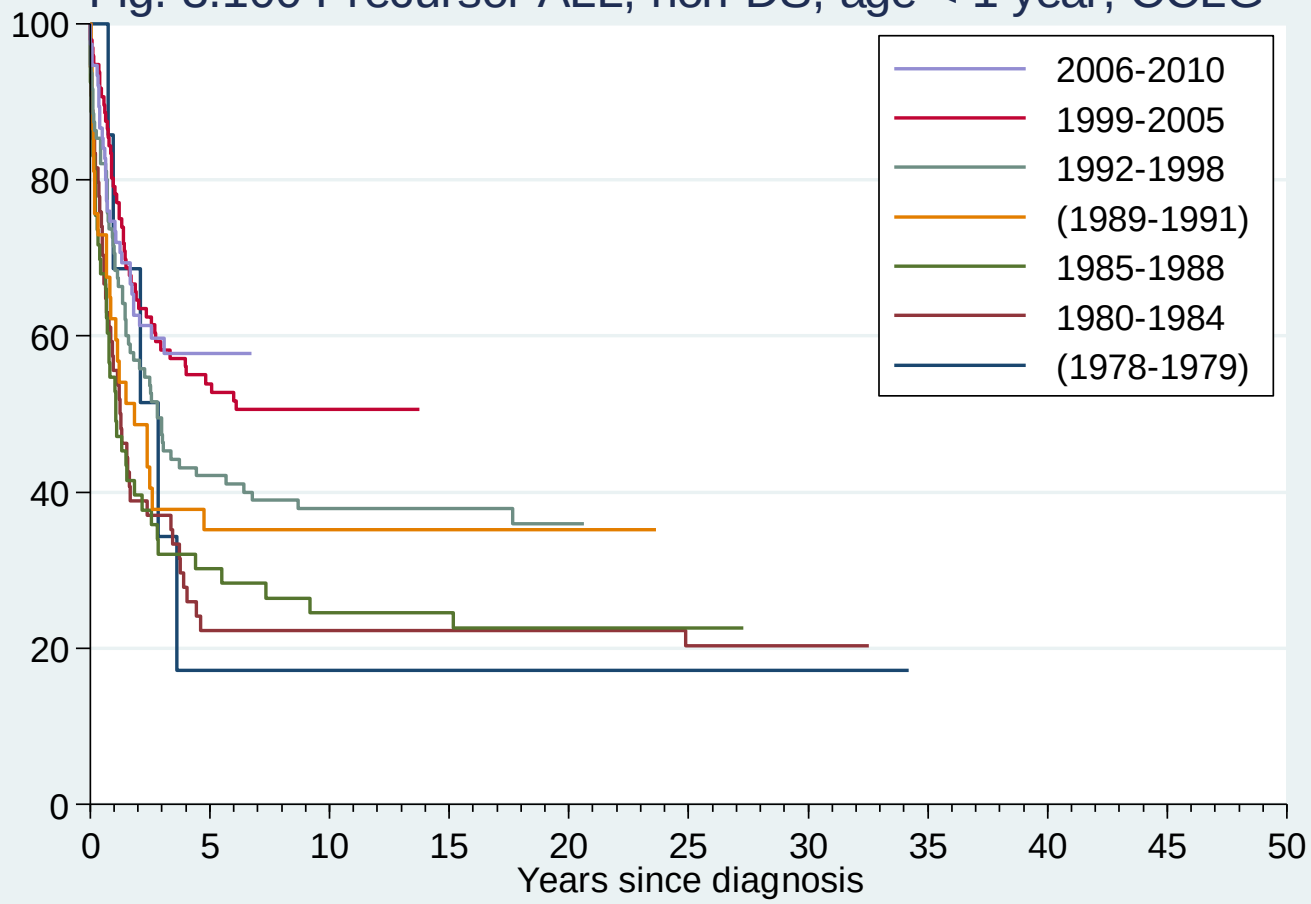


Fig. 3.101 Precursor ALL, non-DS, age 1-14 years, CCLG

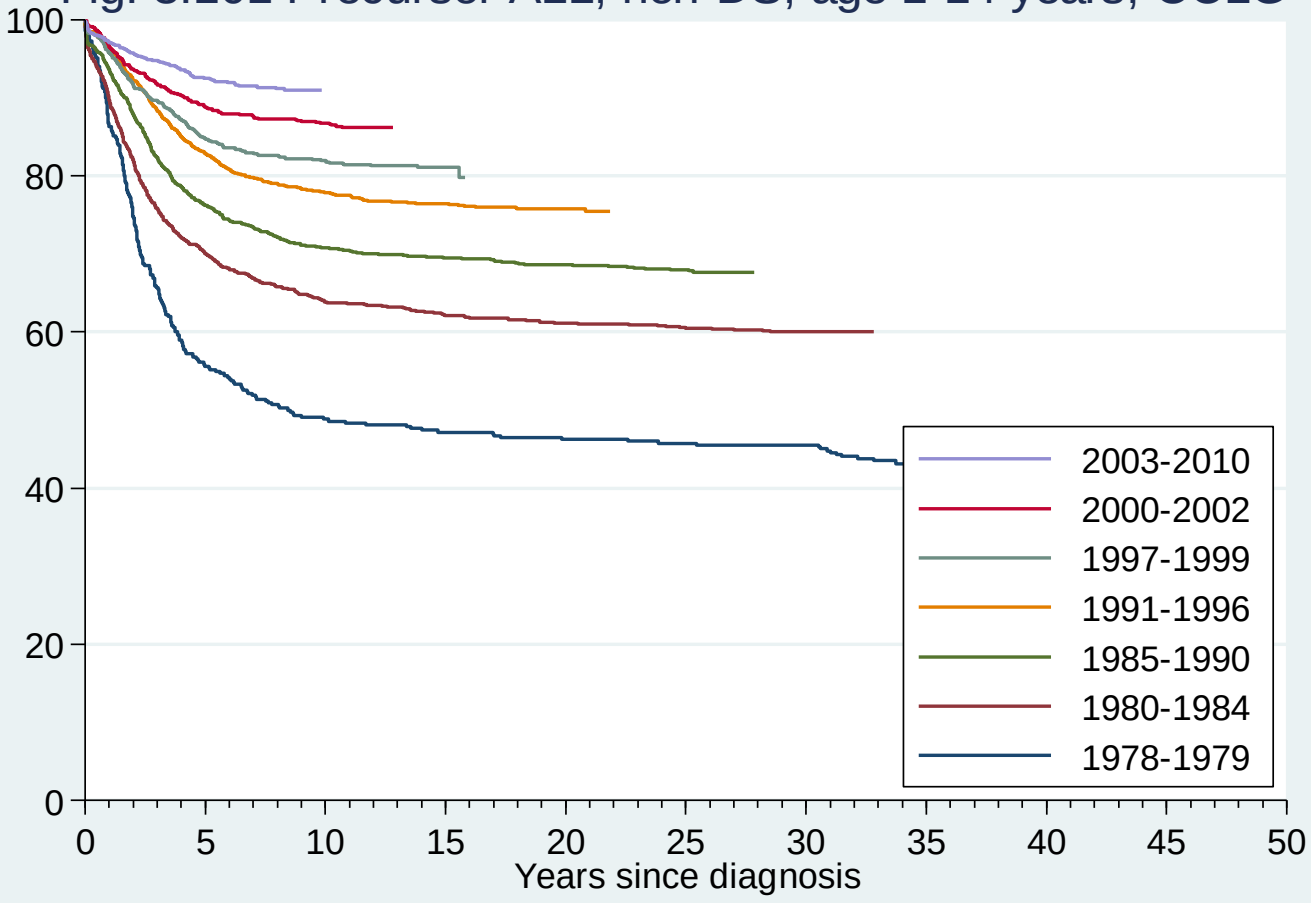


Fig. 3.102 Precursor ALL, Down syndrome, CCLG

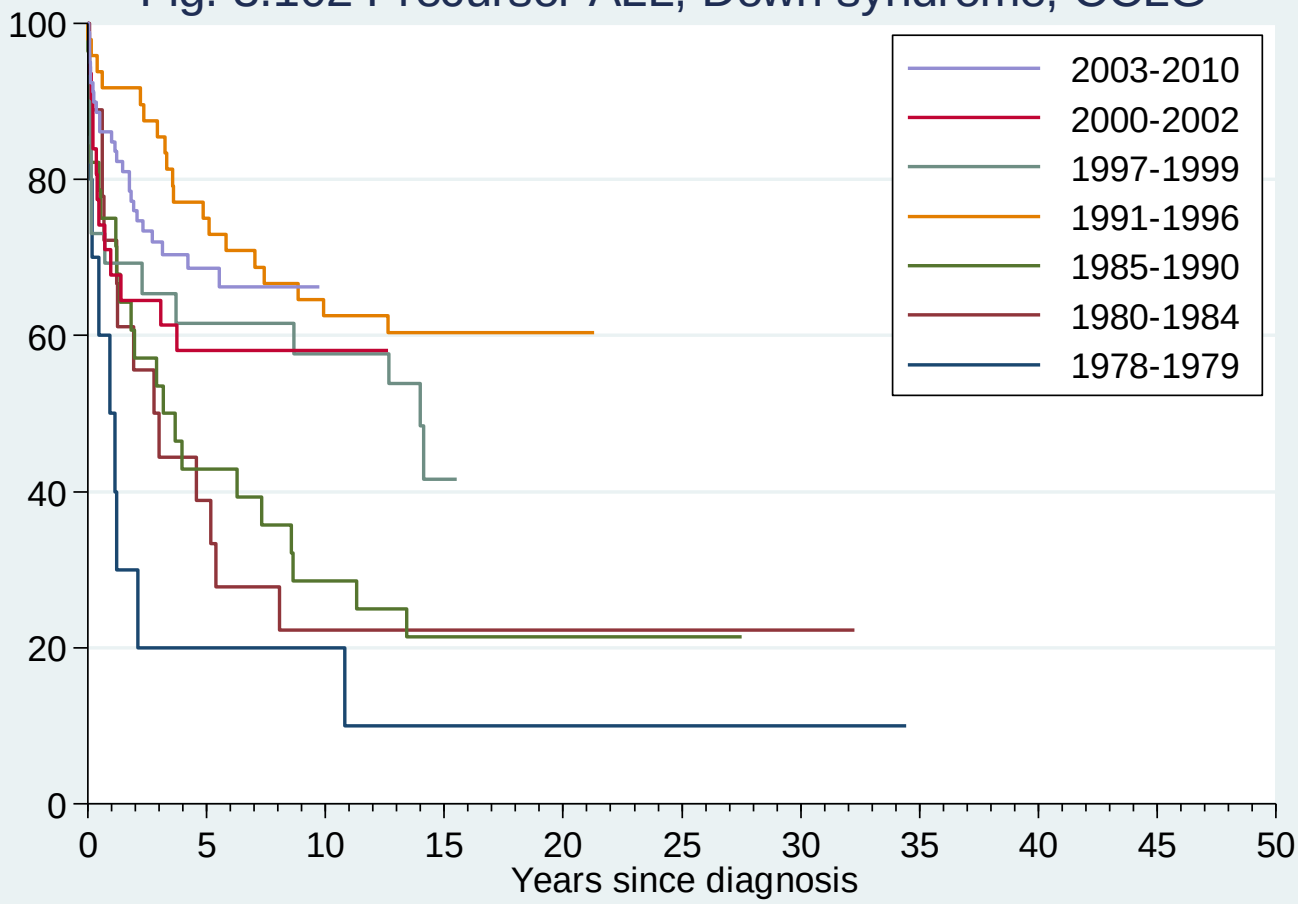


Fig. 3.103 Mature B-cell leukaemia, CCLG

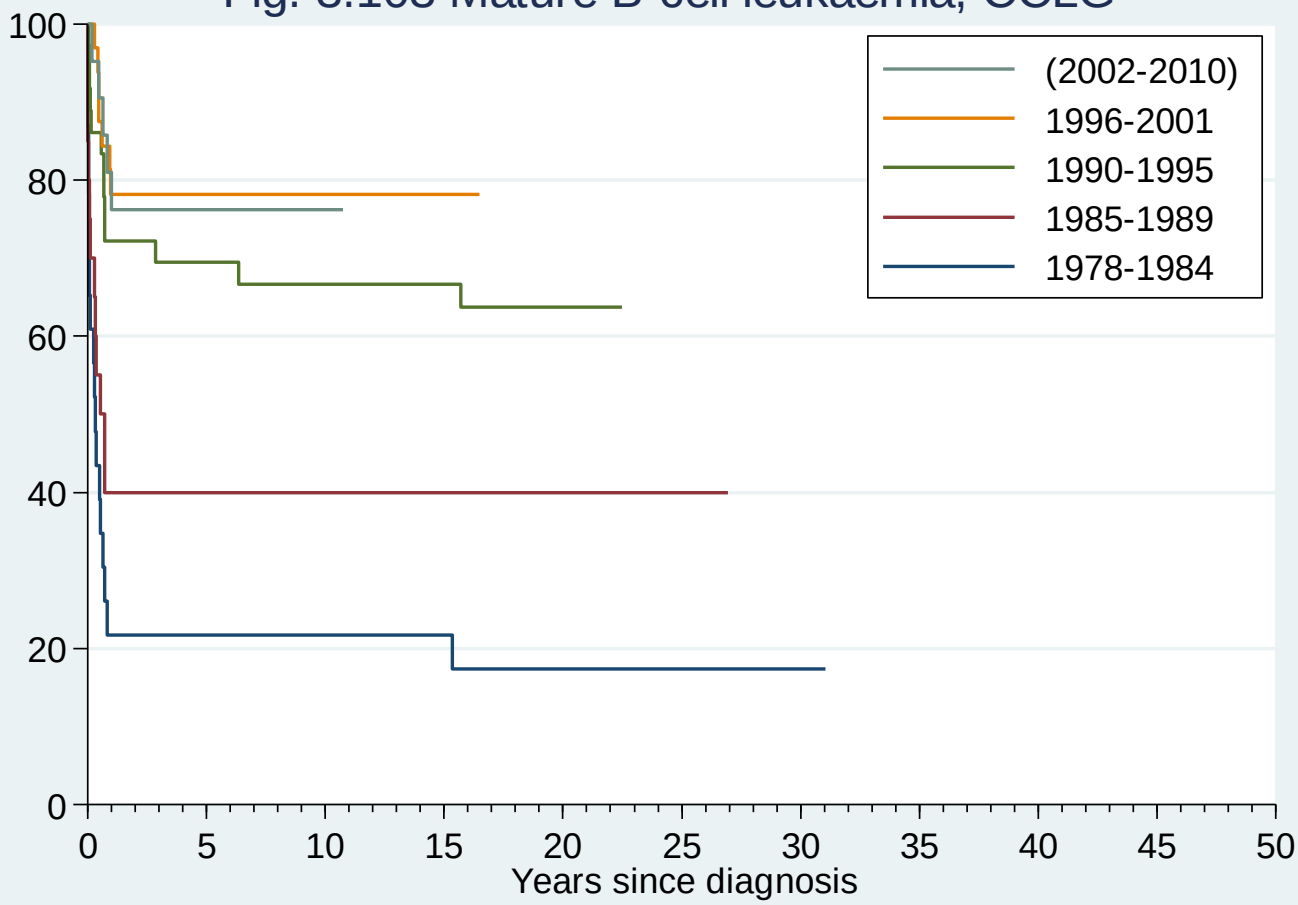


Fig. 3.104 Acute myeloid leukaemia, non-DS, CCLG

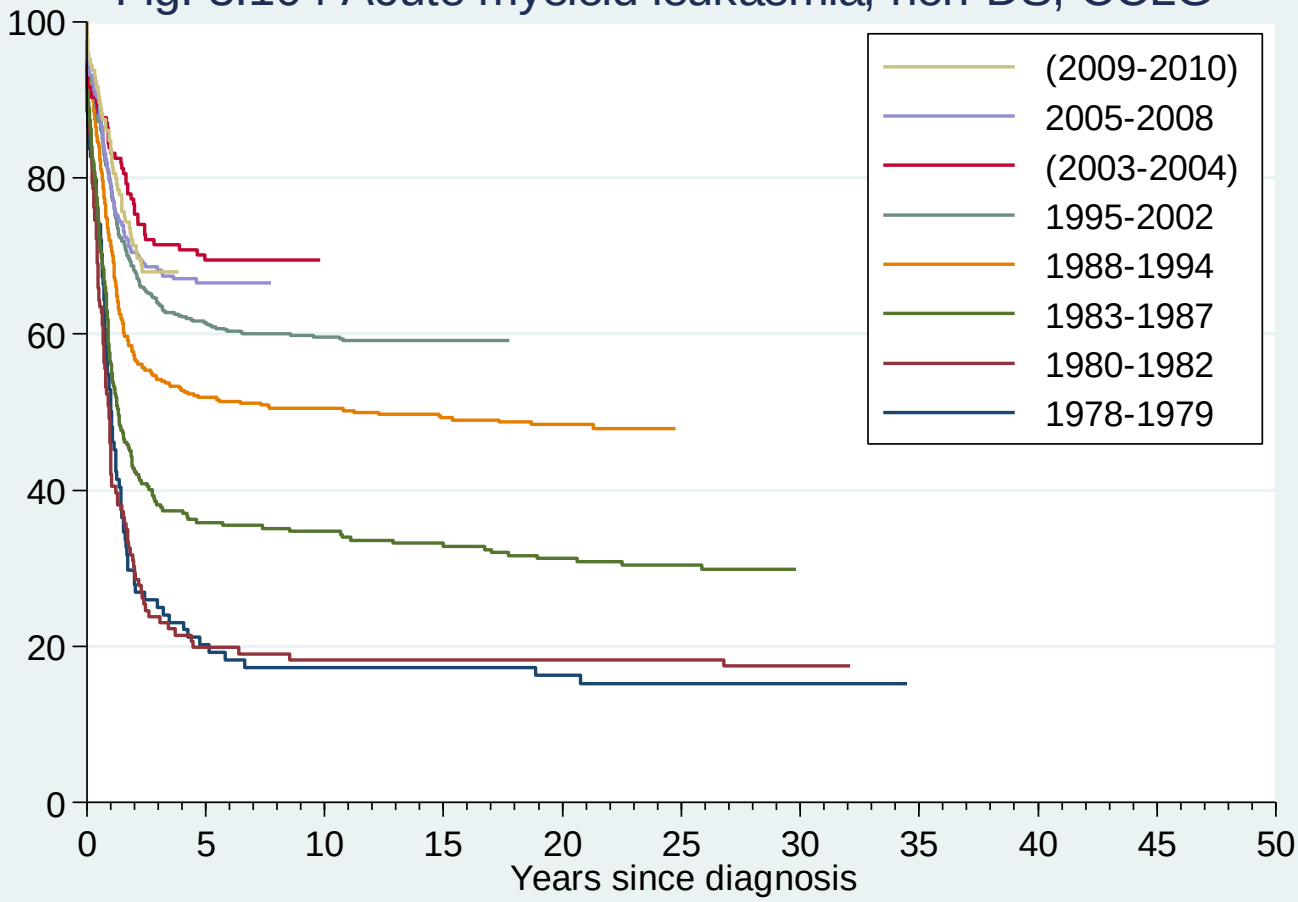


Fig. 3.105 Myeloid leukaemia of Down syndrome, CCLG

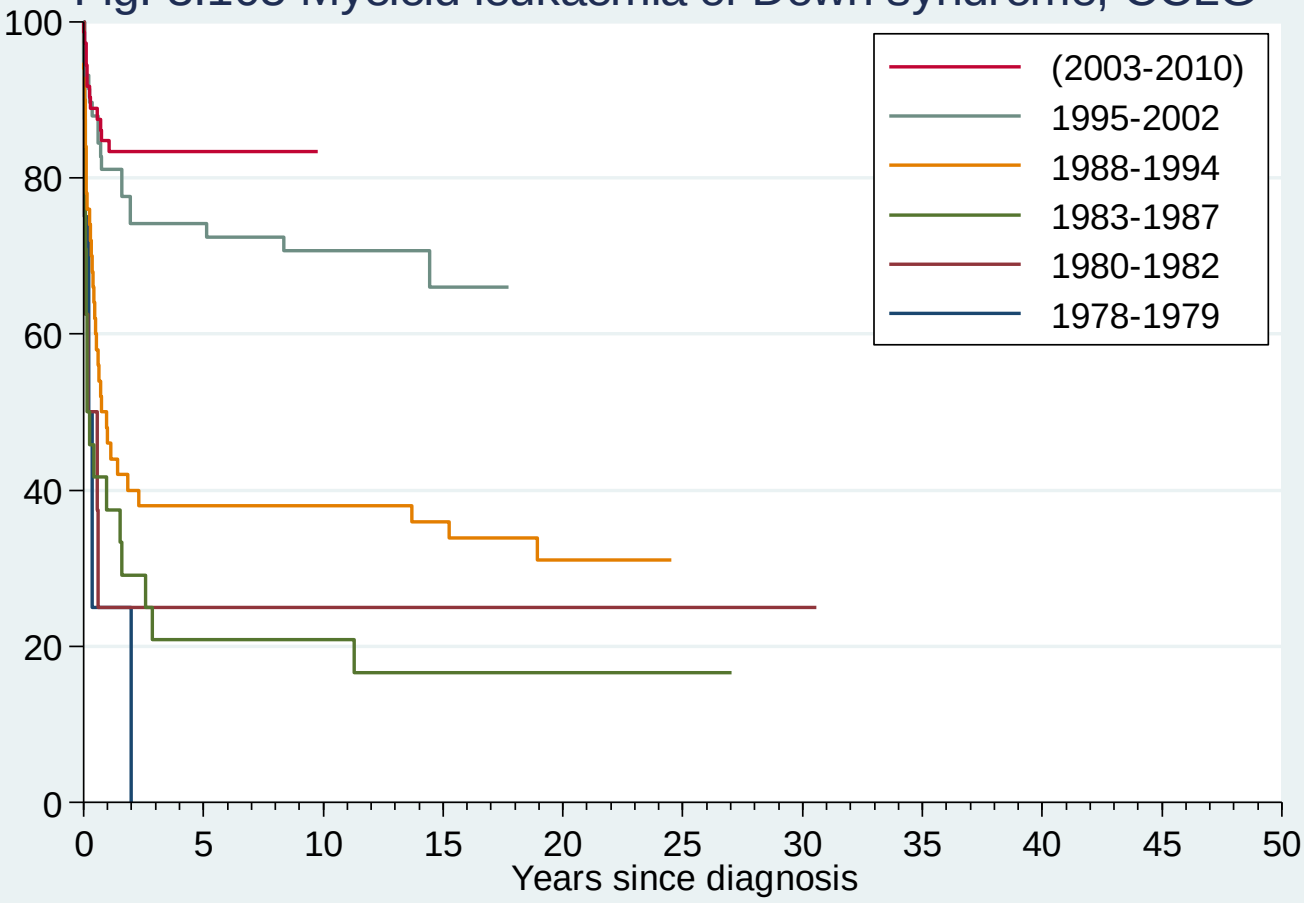


Fig. 3.106 Hodgkin lymphoma, CCLG

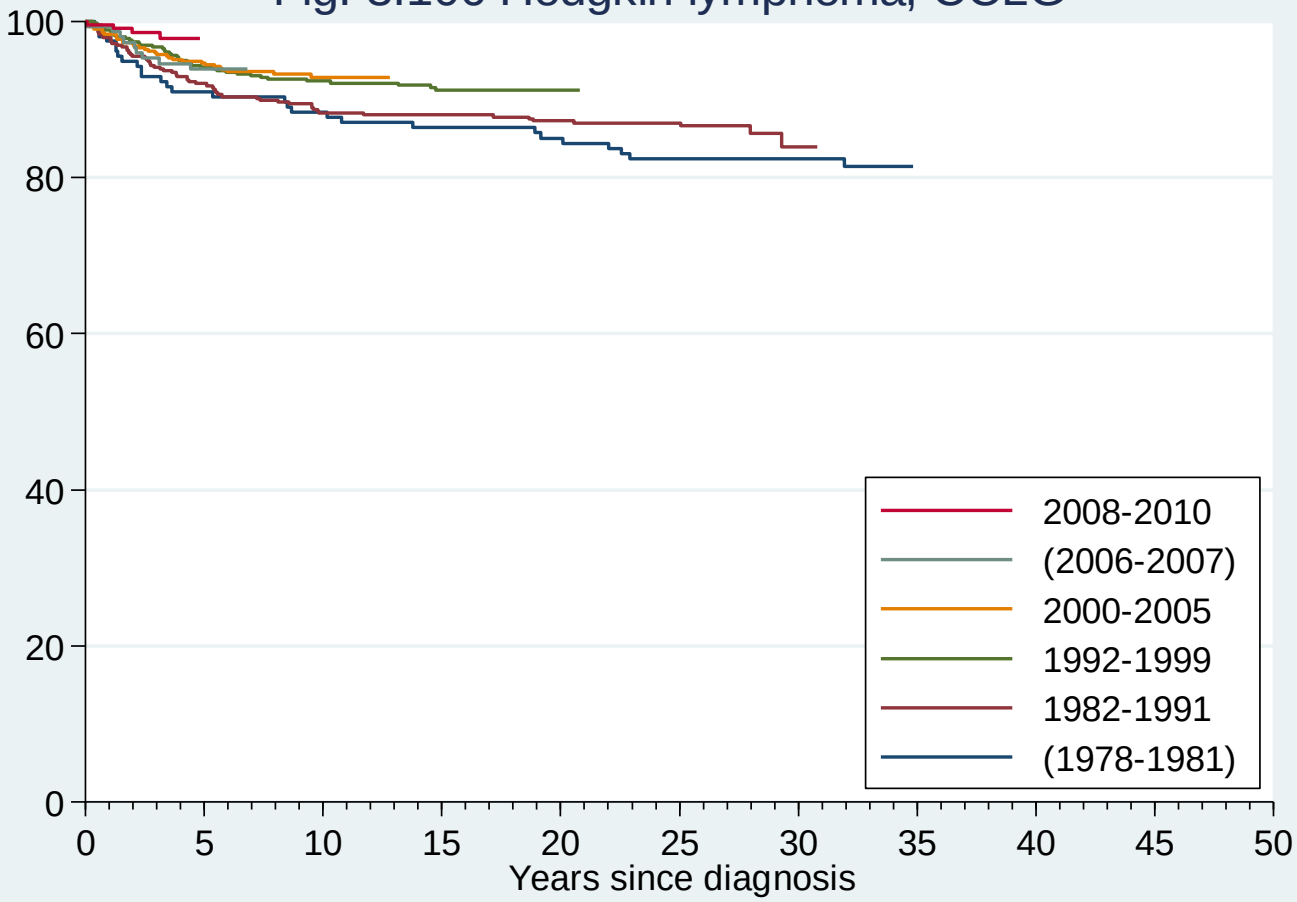


Fig. 3.107 Anaplastic large cell lymphoma, CCLG

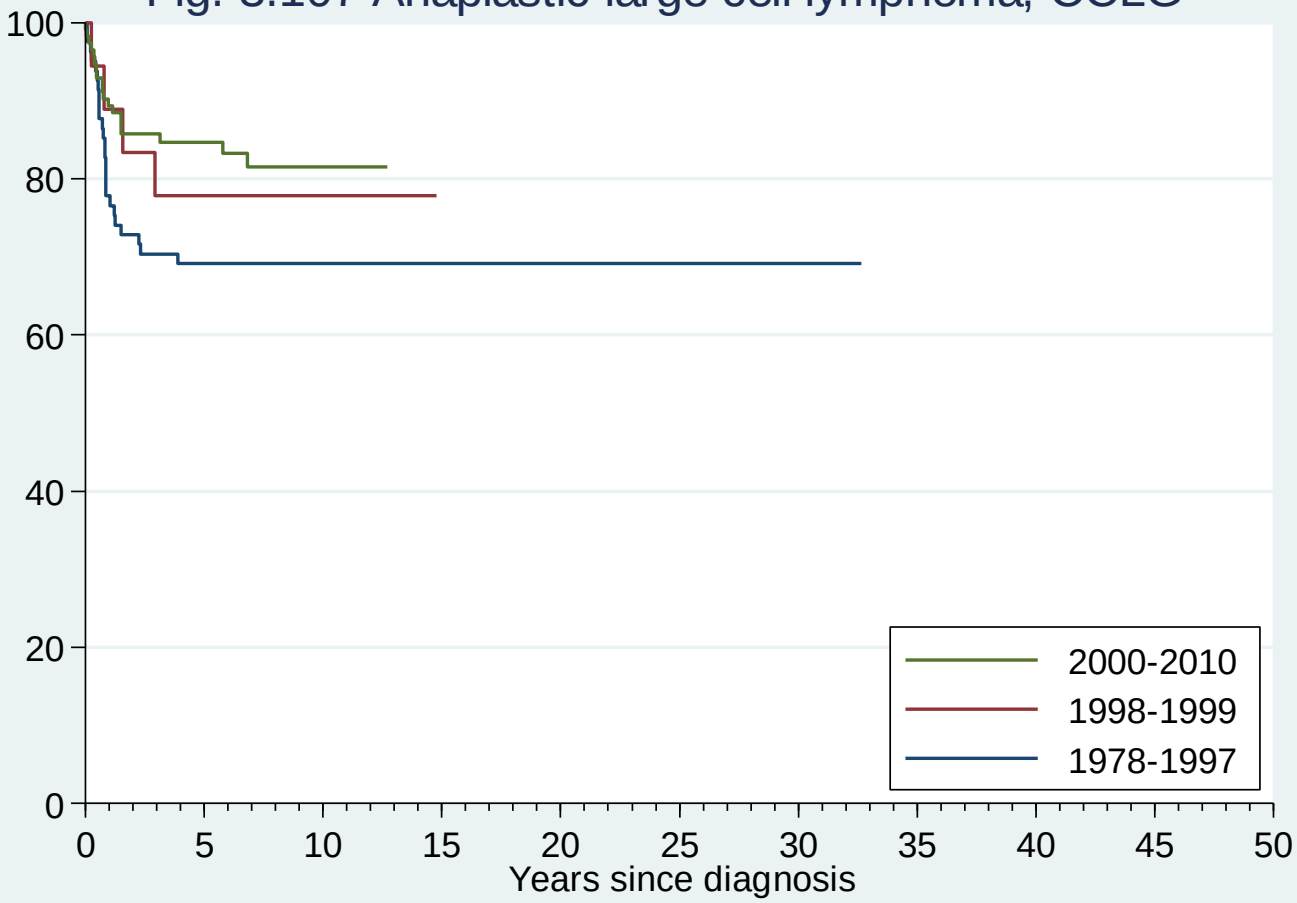


Fig. 3.108 Other T-cell non-Hodgkin lymphoma, CCLG

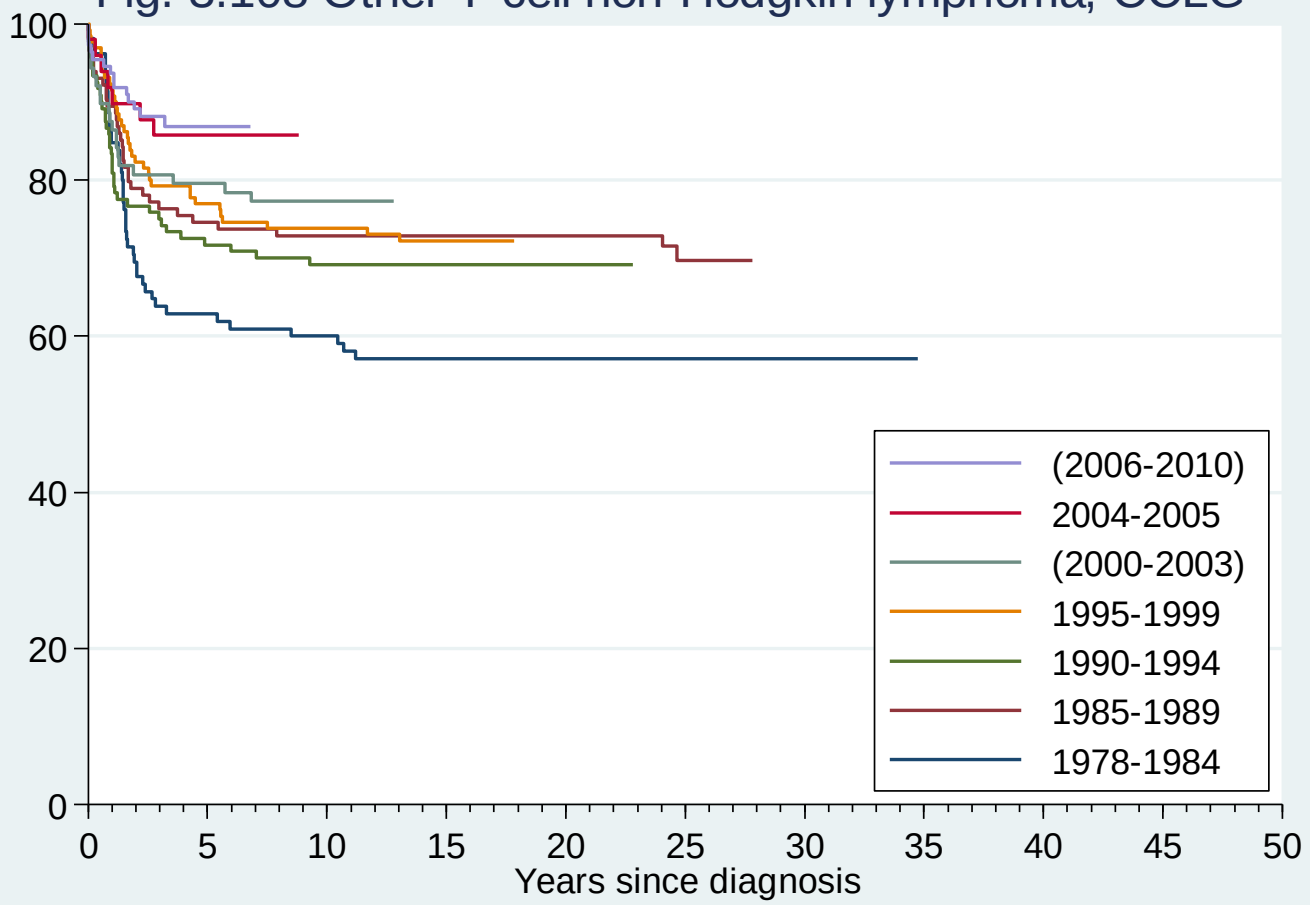


Fig. 3.109 B-cell non-Hodgkin lymphoma, CCLG

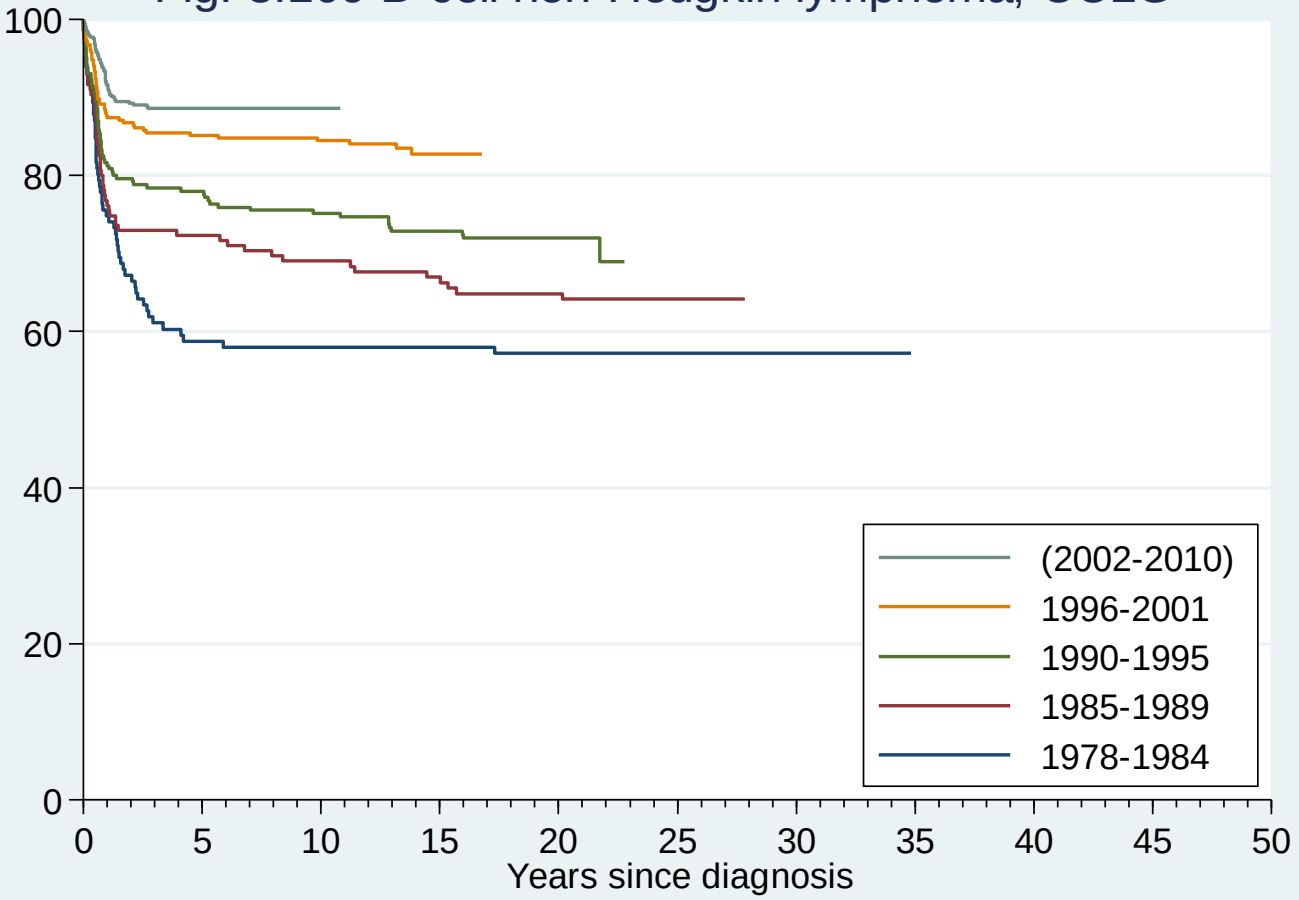


Fig. 3.110 Ependymoma, age 0-2 years, CCLG

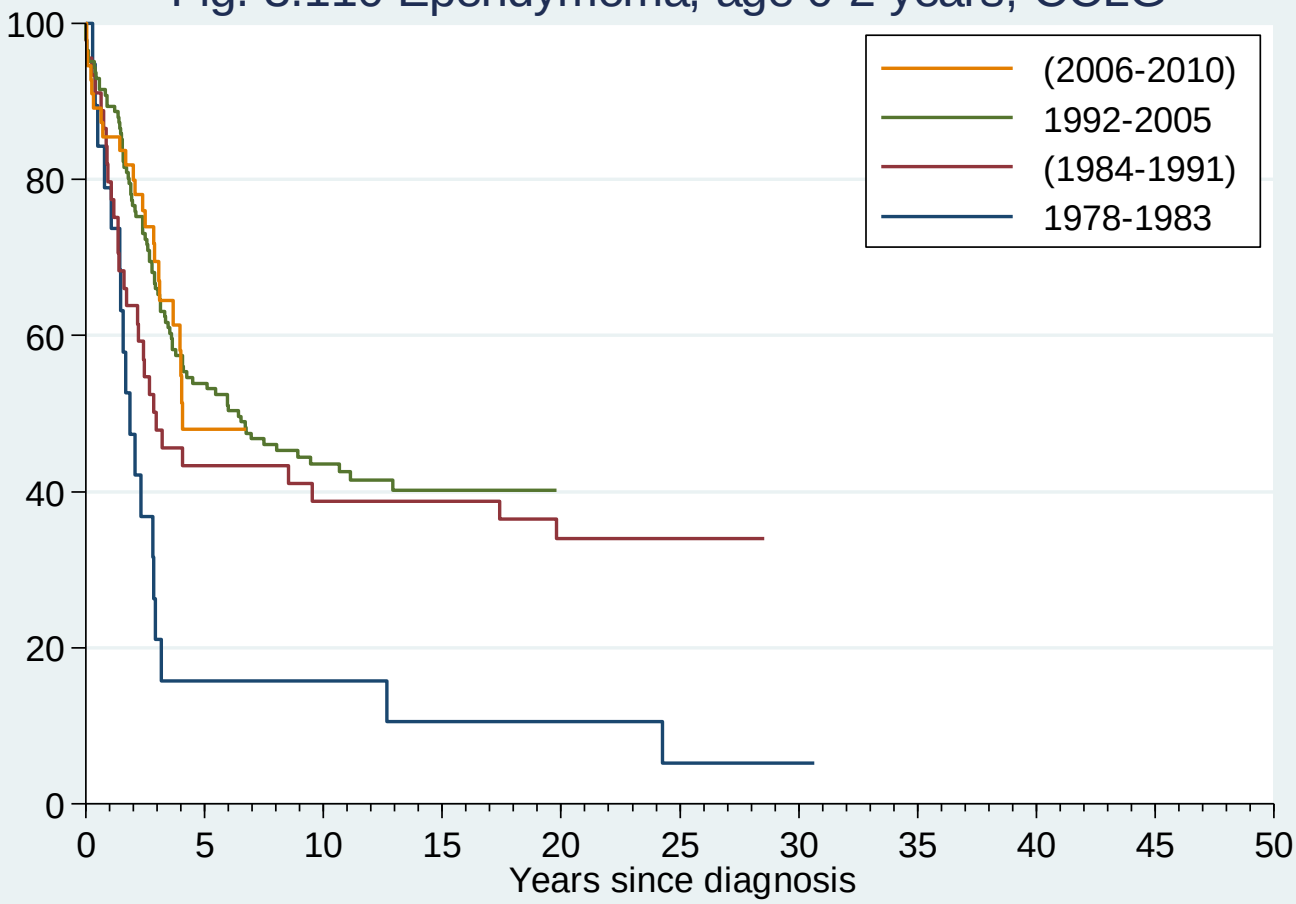


Fig. 3.111 Ependymoma, age 3-14 years, CCLG

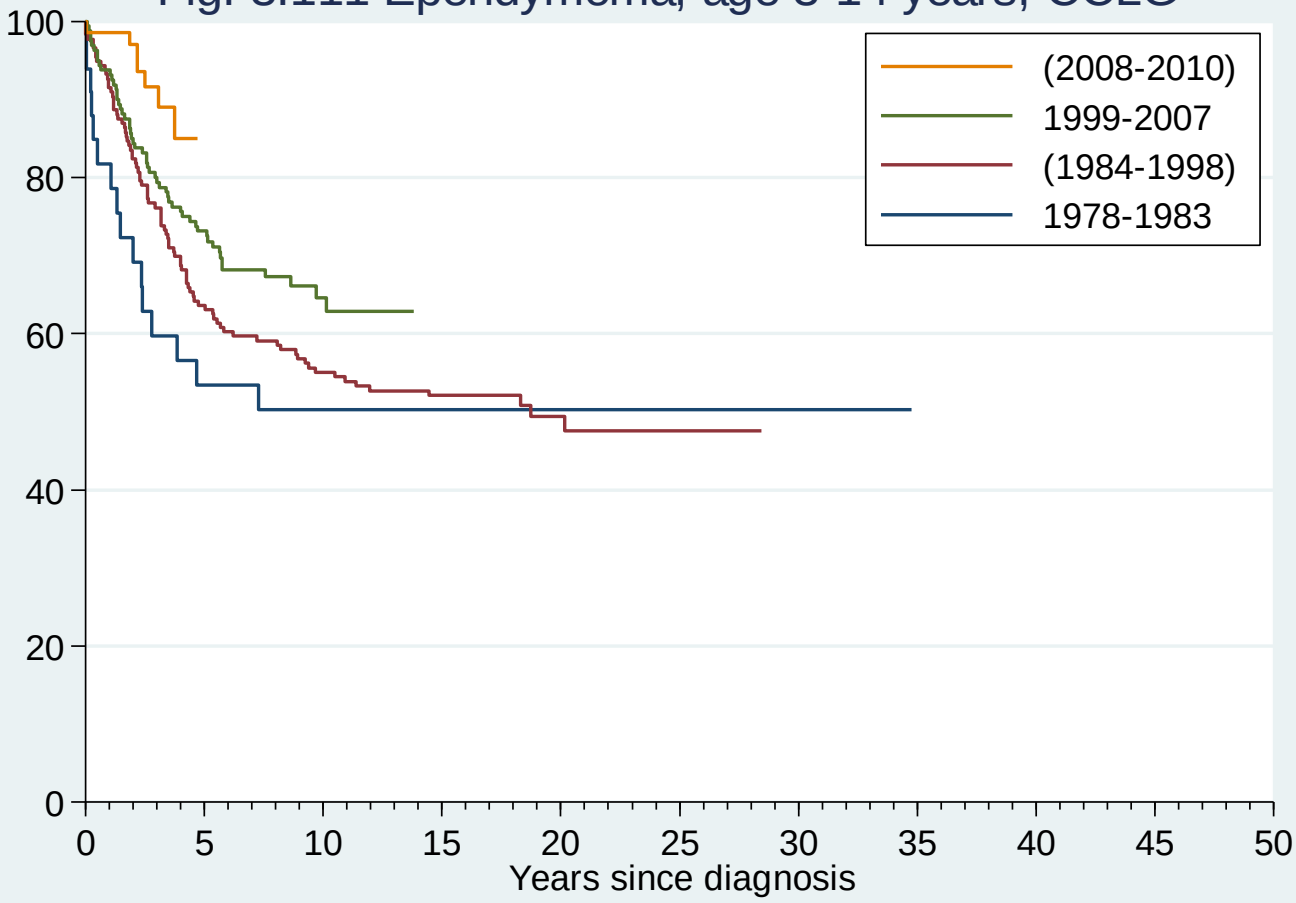


Fig. 3.112 Low-grade astrocytoma, CCLG

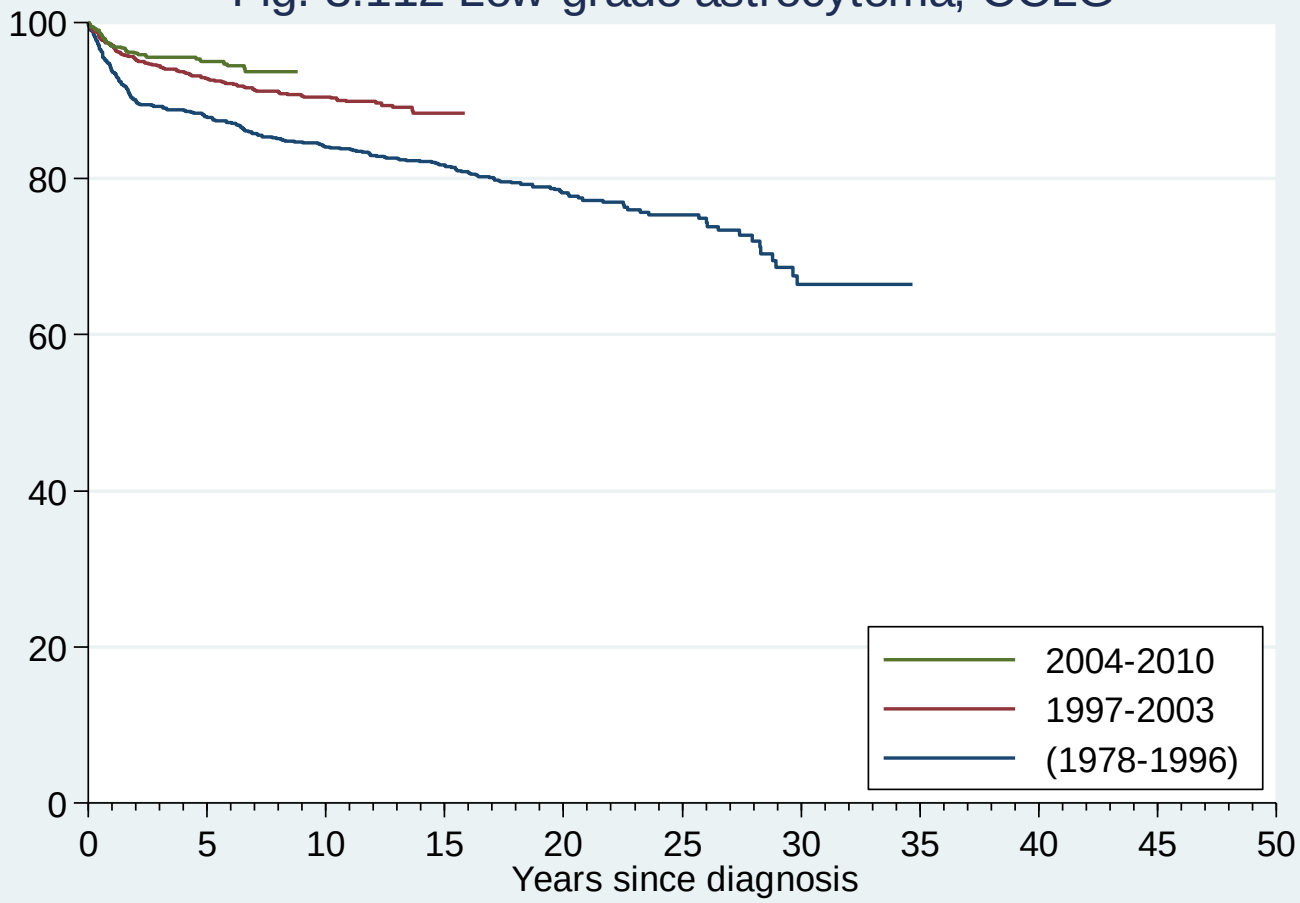


Fig. 3.113 Embryonal CNS tumours, age 0-2 years, CCLG

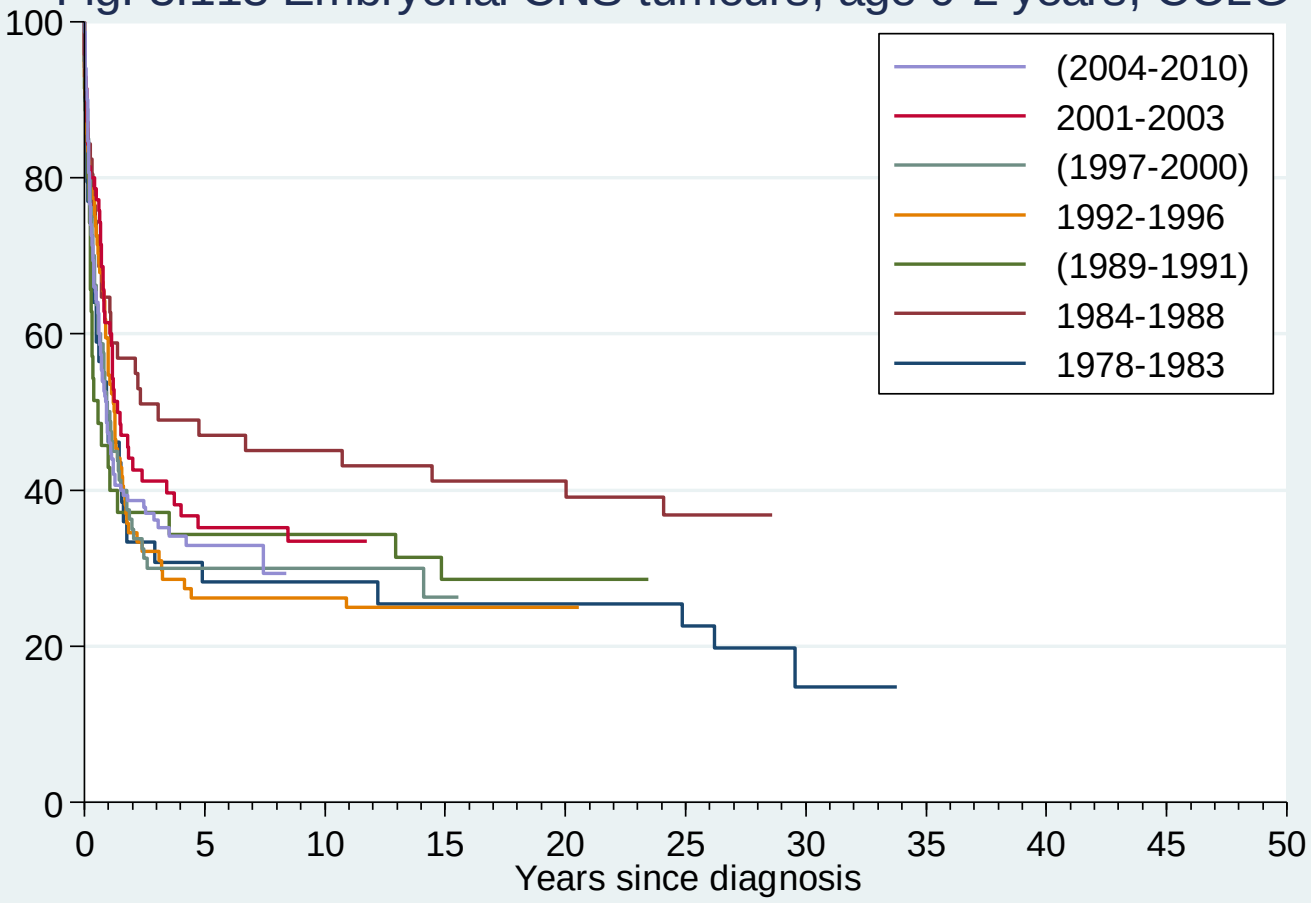


Fig. 3.114 Embryonal CNS tumours, age 3-14 years, CCLG

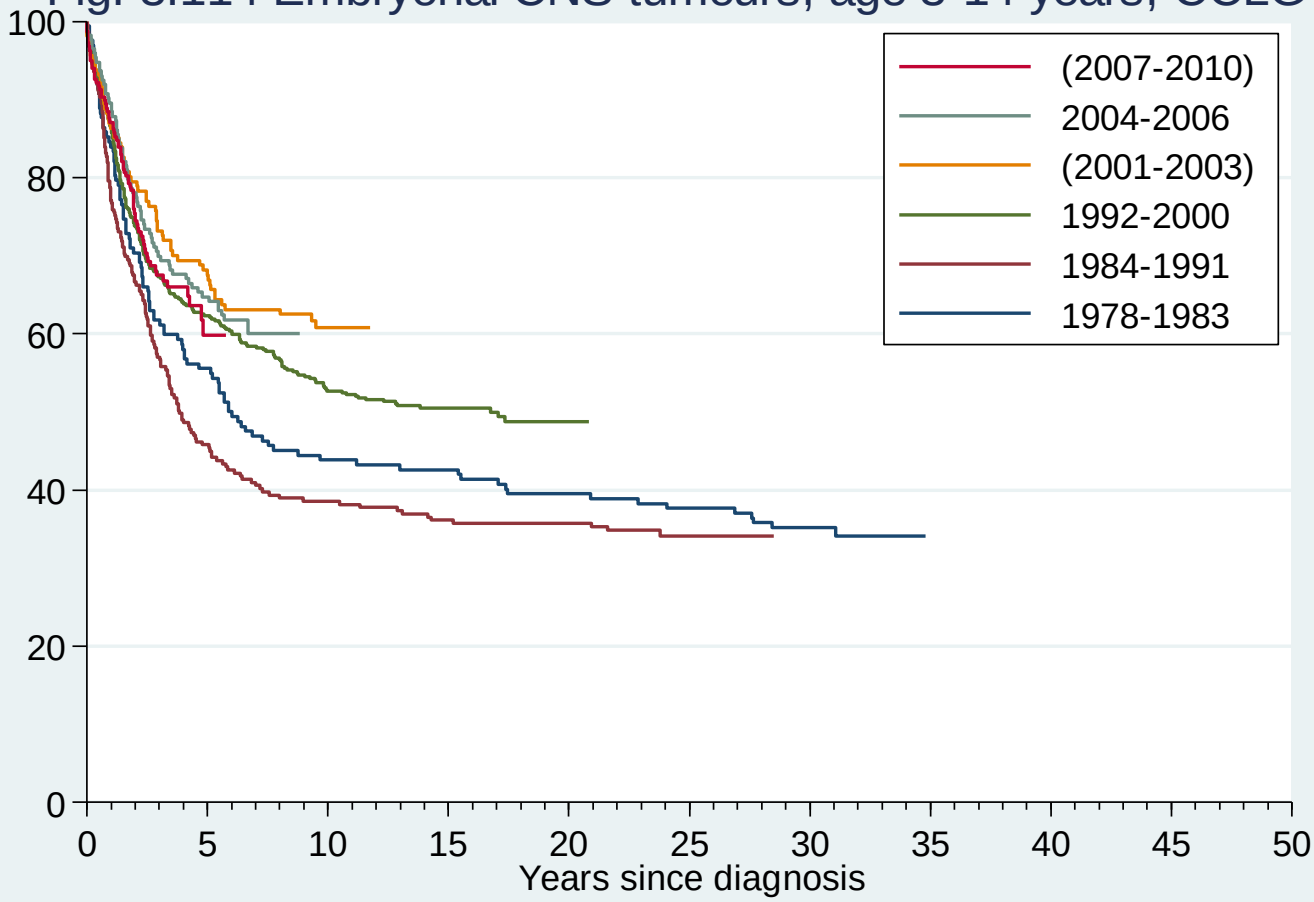


Fig. 3.115 Neuroblastoma, age < 1 year, CCLG

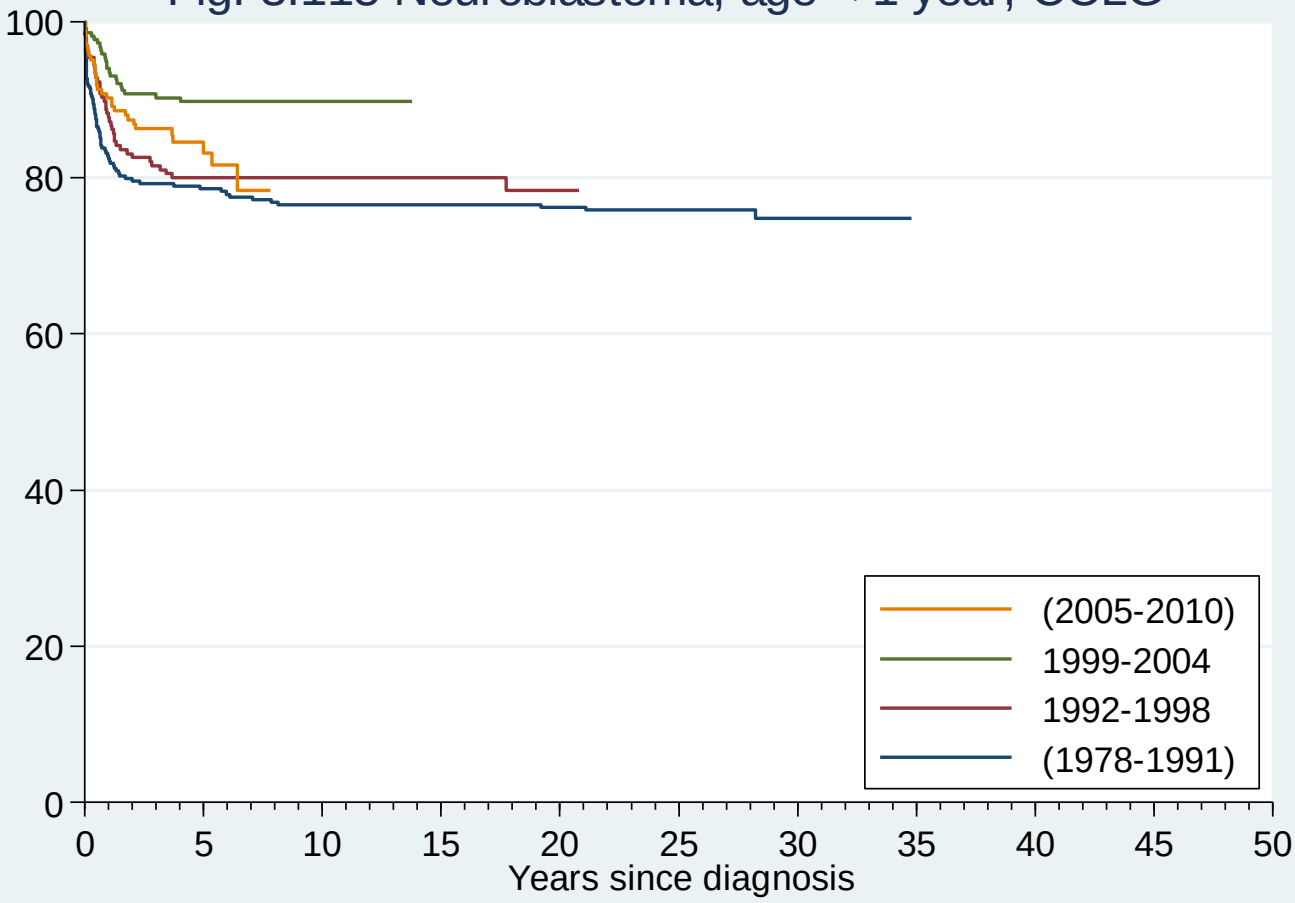


Fig. 3.116 Neuroblastoma, age 1-14 years, CCLG

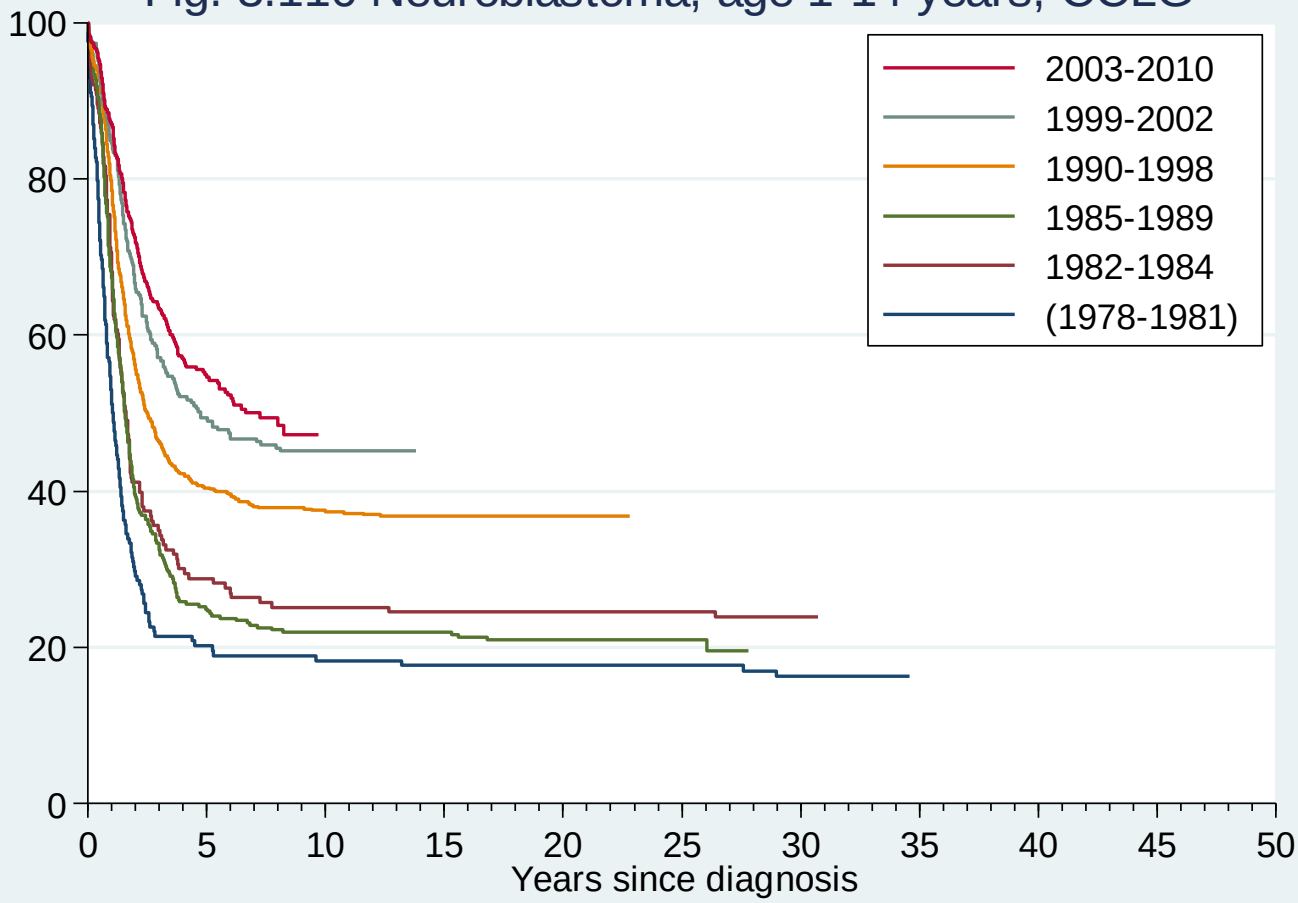


Fig. 3.117 Wilms tumour, CCLG

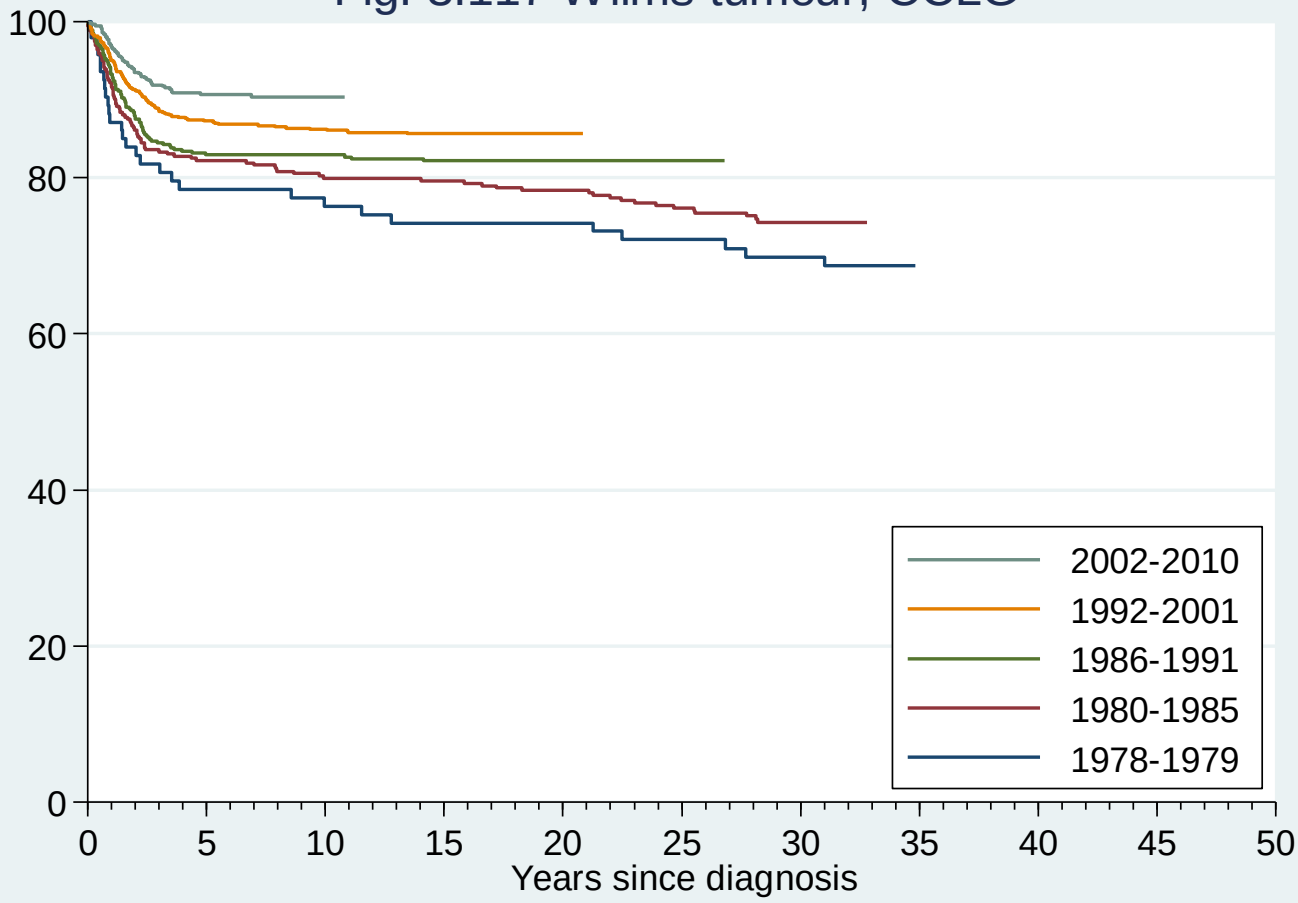


Fig. 3.118 Rhabdoid renal tumour, CCLG

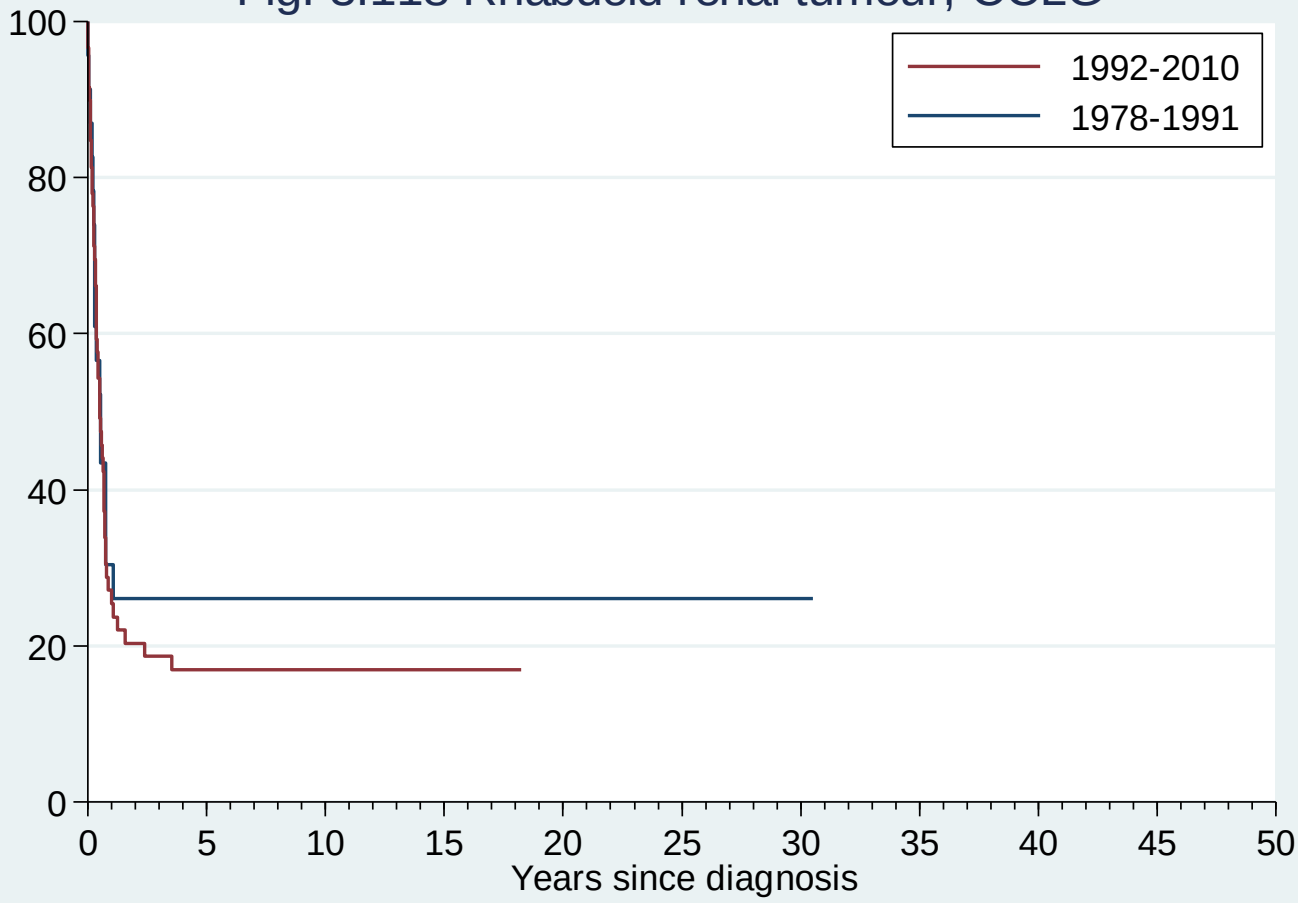


Fig. 3.119 Renal clear-cell sarcoma, CCLG

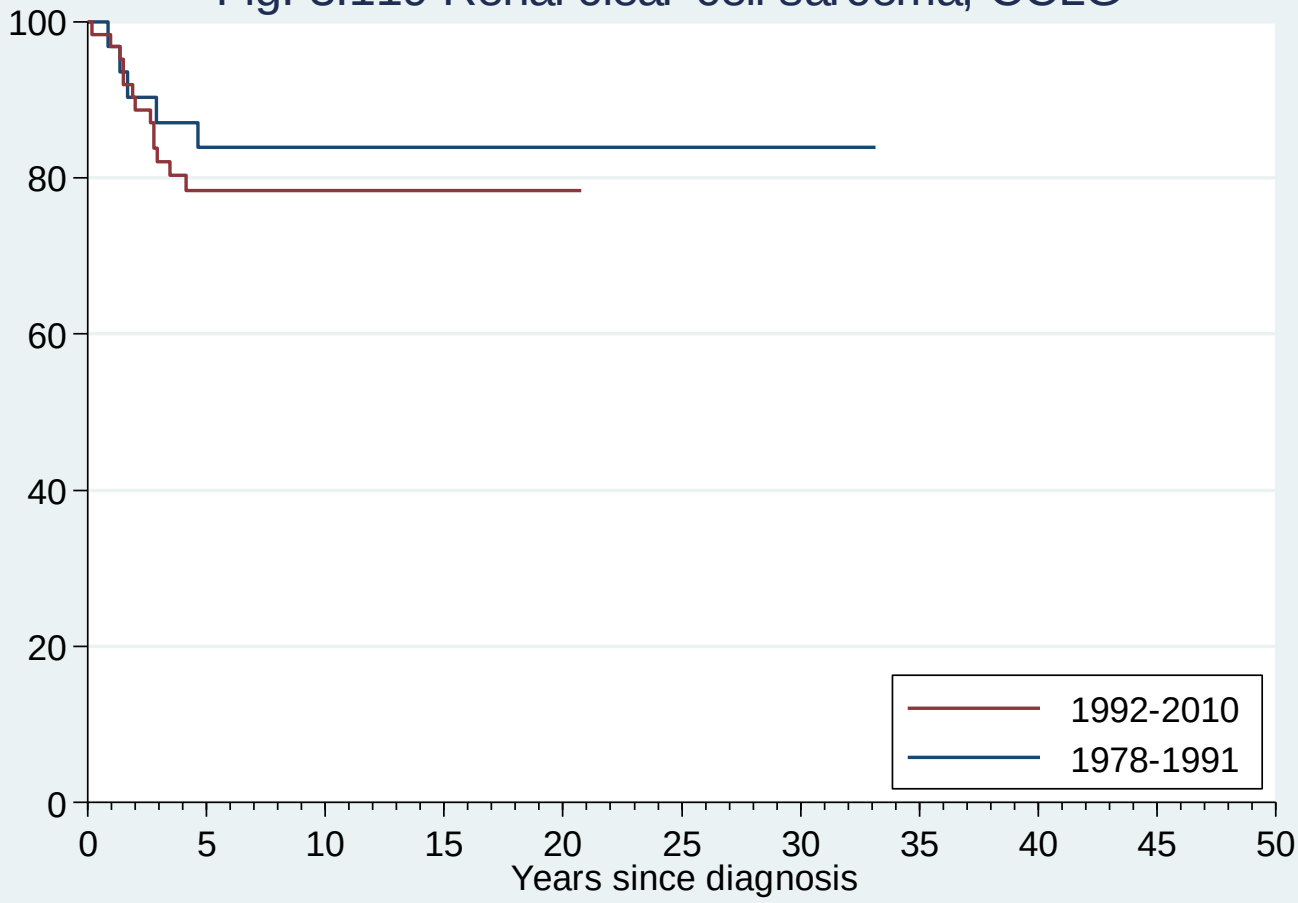


Fig. 3.120 Hepatoblastoma, CCLG

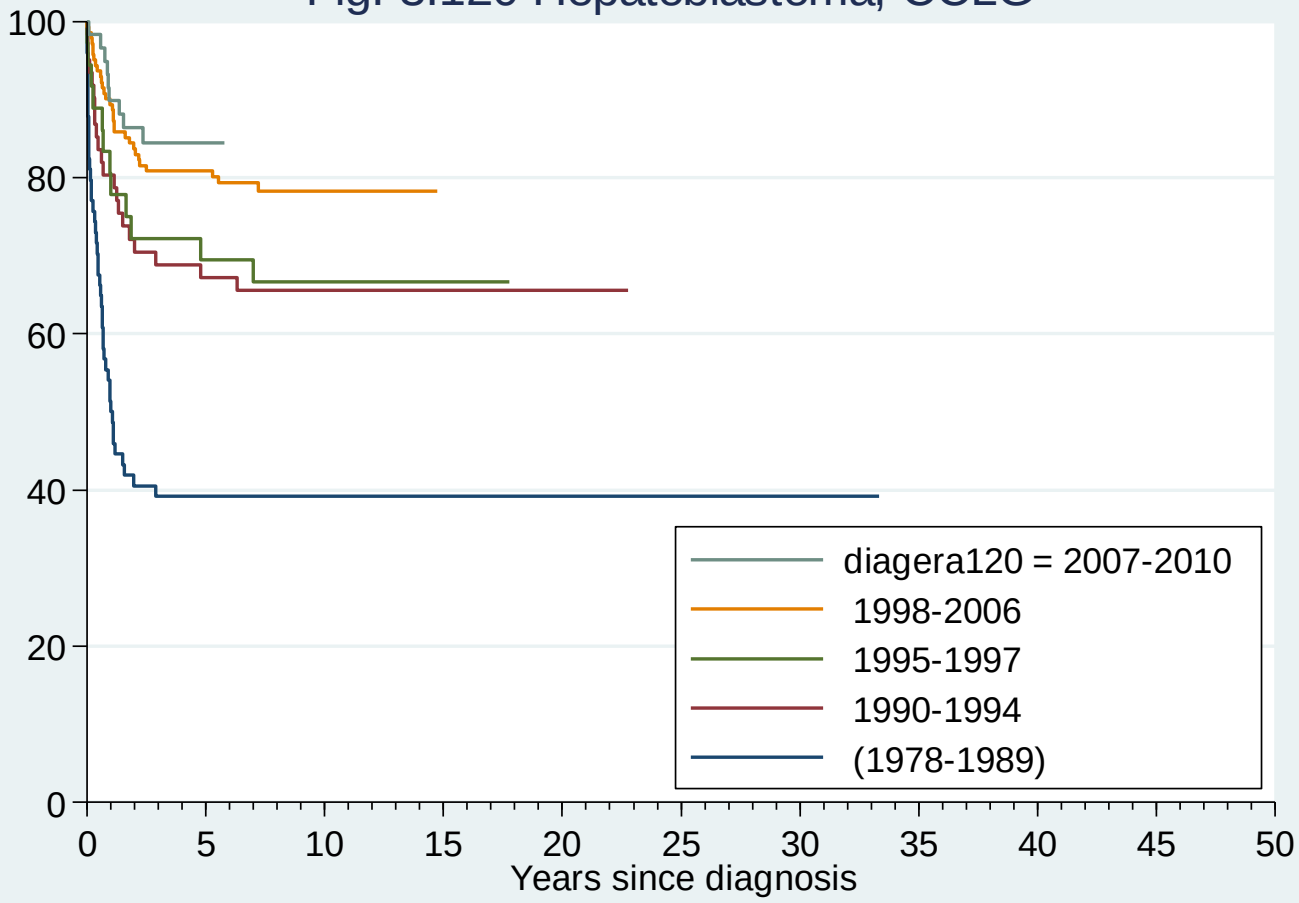


Fig. 3.121 Hepatic carcinoma, CCLG

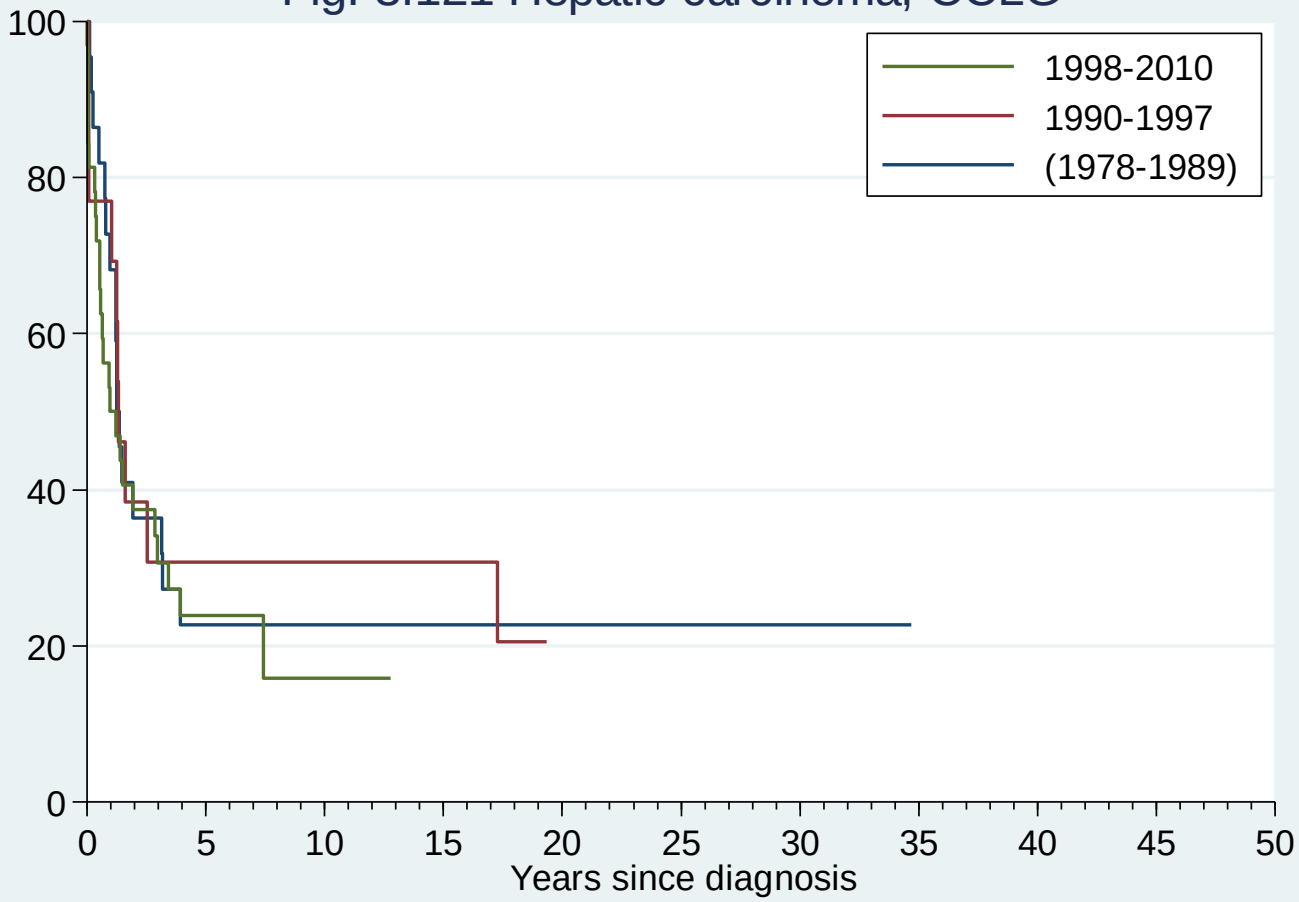


Fig. 3.122 Osteosarcoma, CCLG

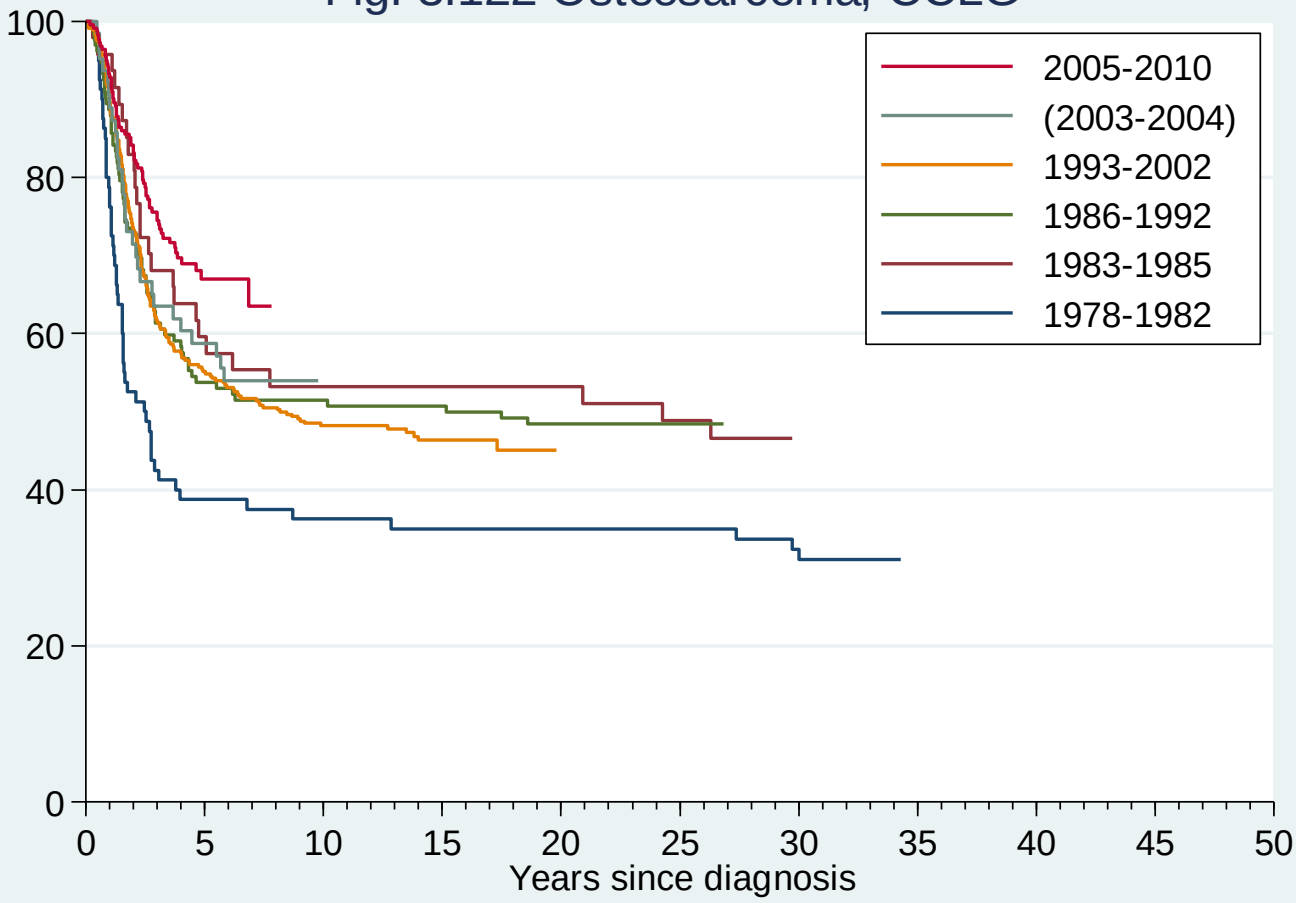


Fig. 3.123 Ewing sarcoma of bone, CCLG

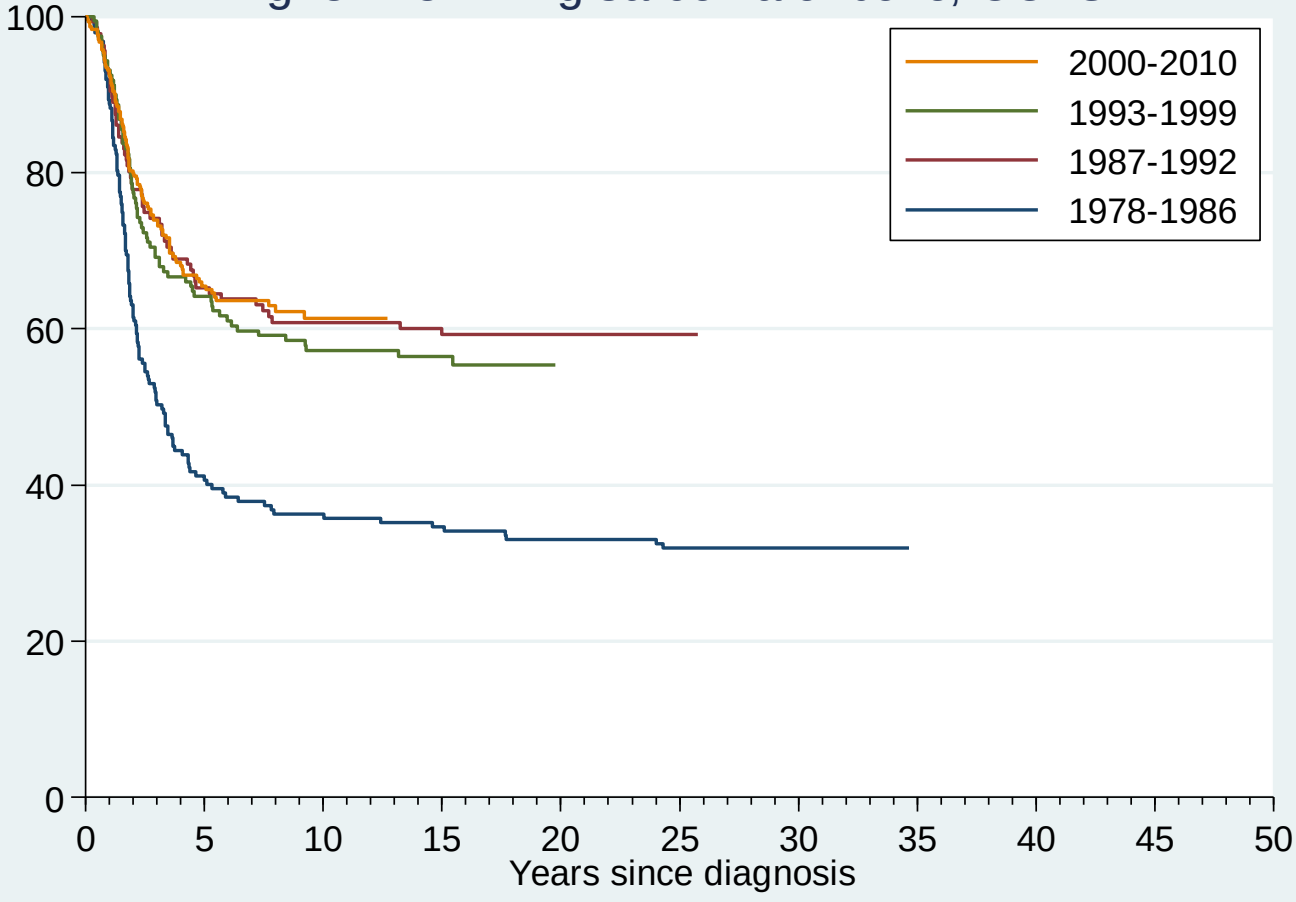


Fig. 3.124 Rhabdomyosarcoma, CCLG

Fig. 3.125 Intracranial and intraspinal germinoma, CCLG

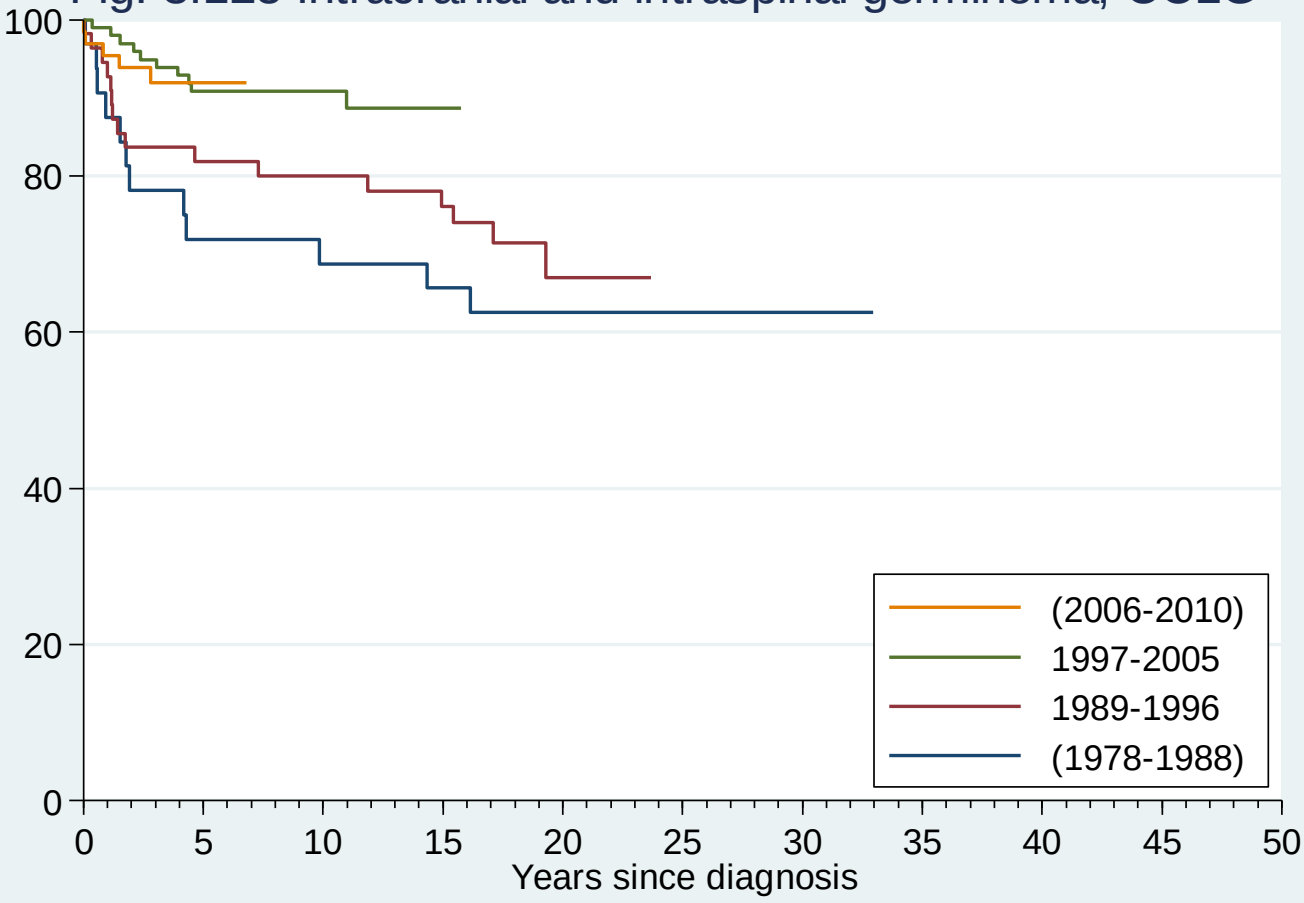


Fig. 3.126 Other intracranial and intraspinal germ-cell, CCLG

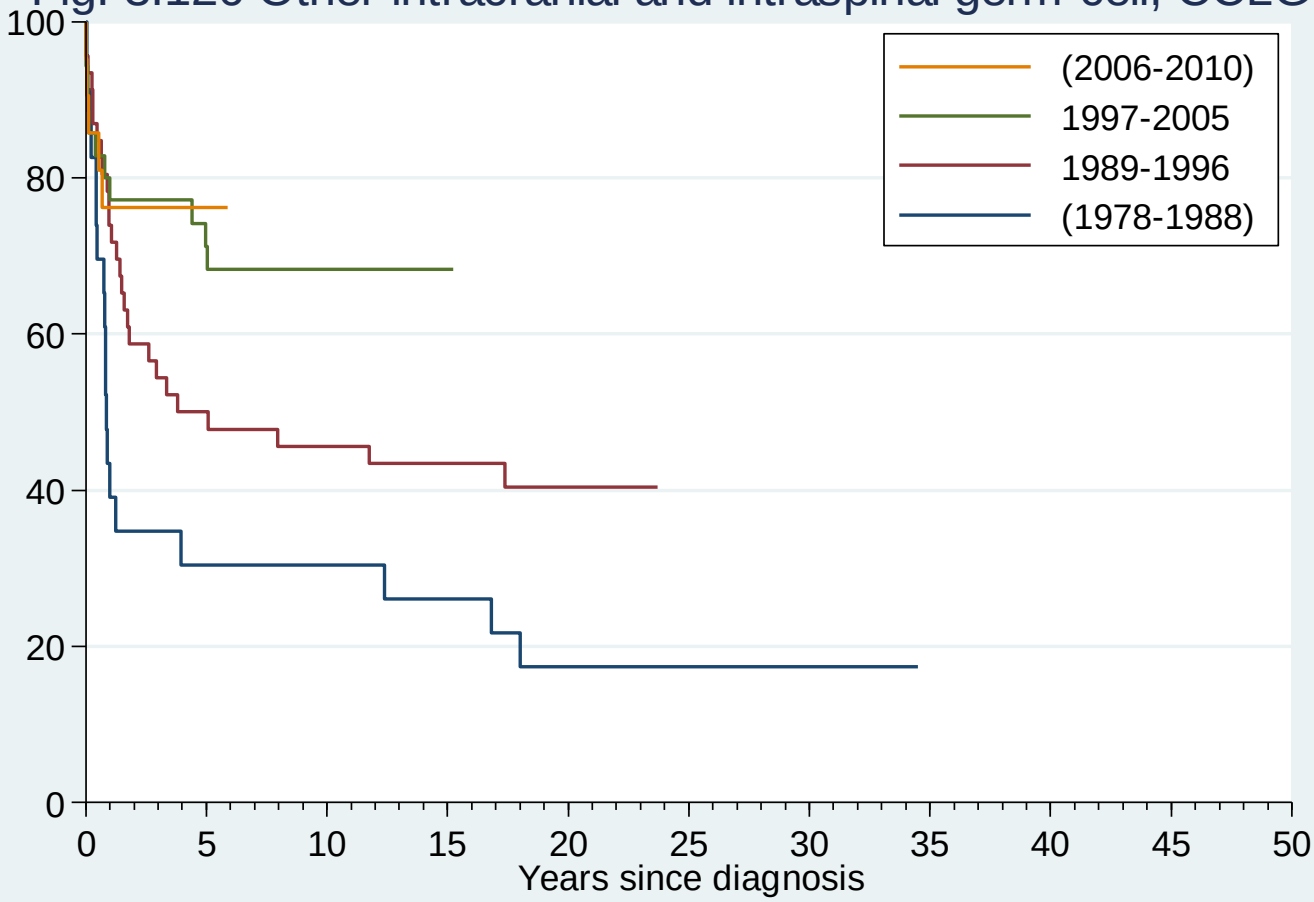


Fig. 3.127 Other malignant extragonadal germ-cell, CCLG

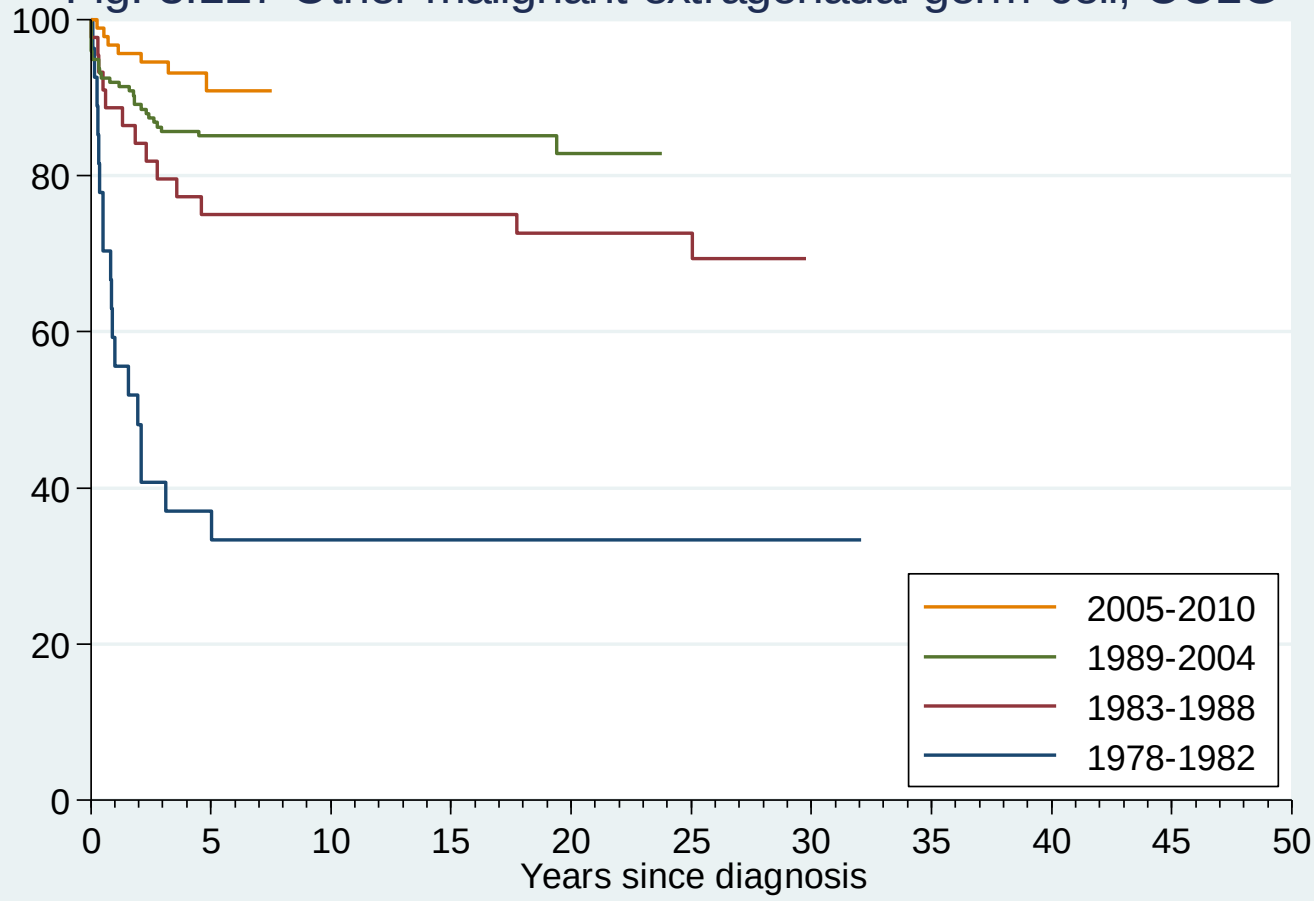


Fig. 3.128 Testicular malignant germ-cell tumours, CCLG

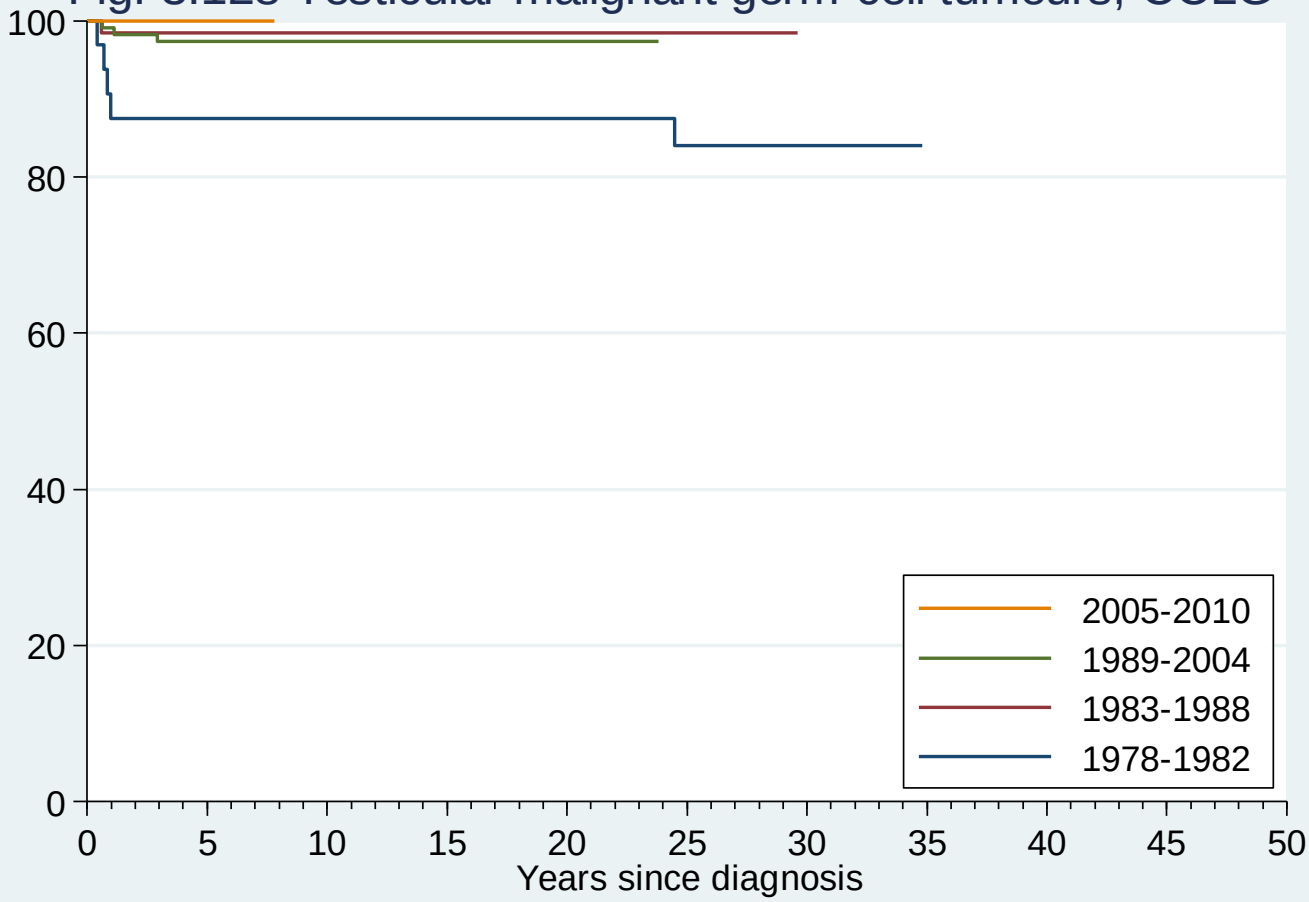


Fig. 3.129 Ovarian malignant germ-cell tumours, CCLG

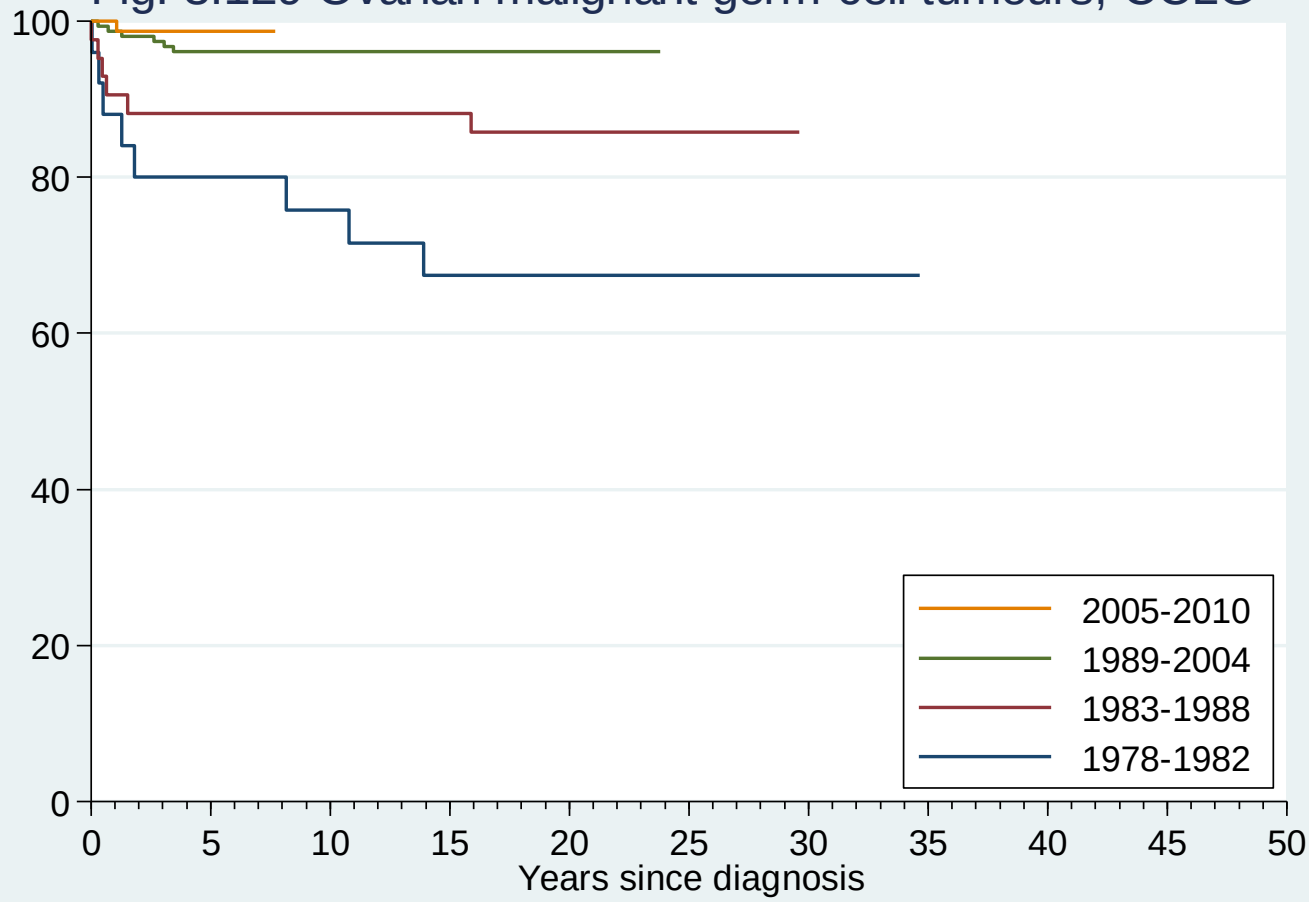


Fig. 3.130 Langerhans cell histiocytosis, sing. system, CCLG

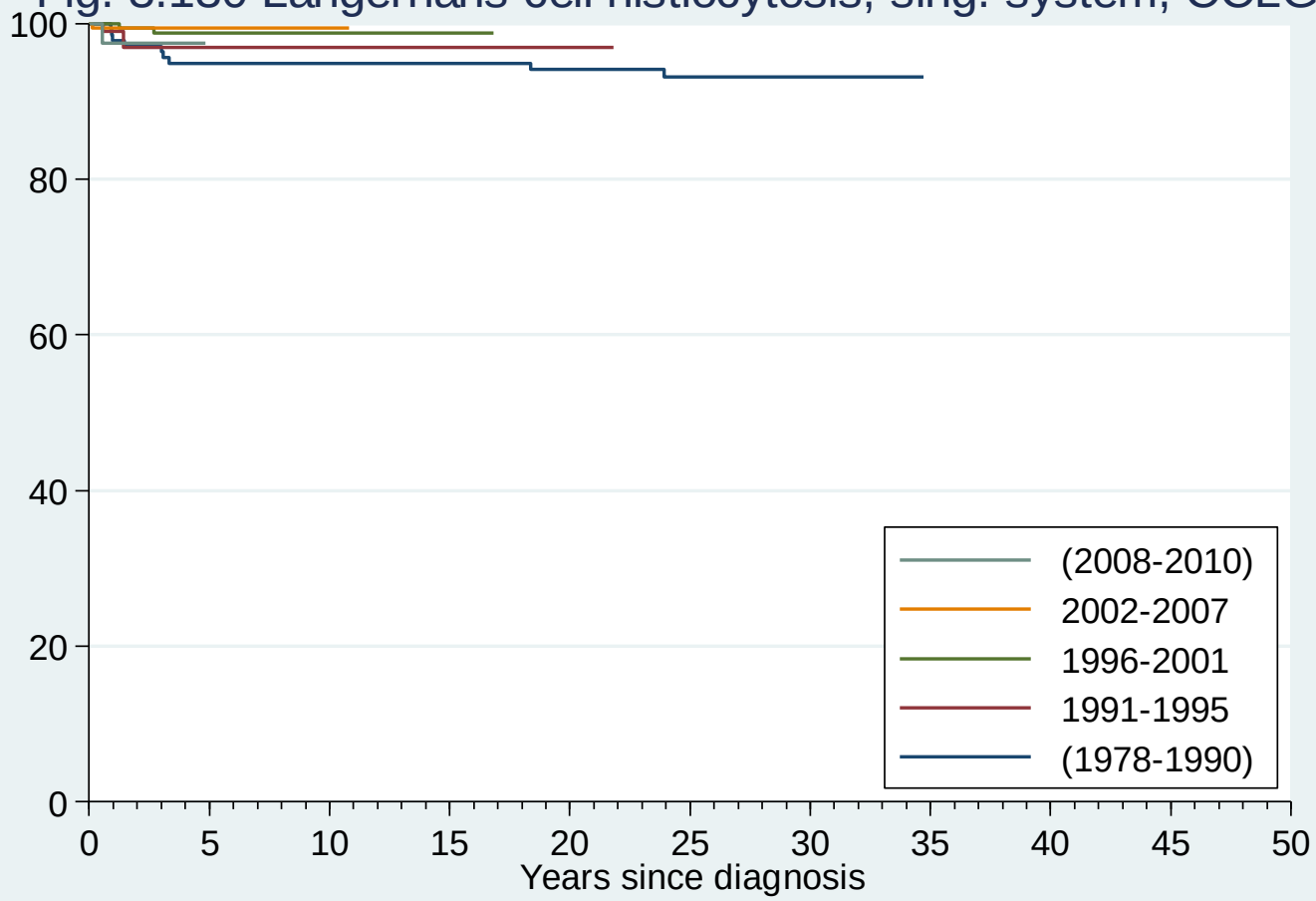
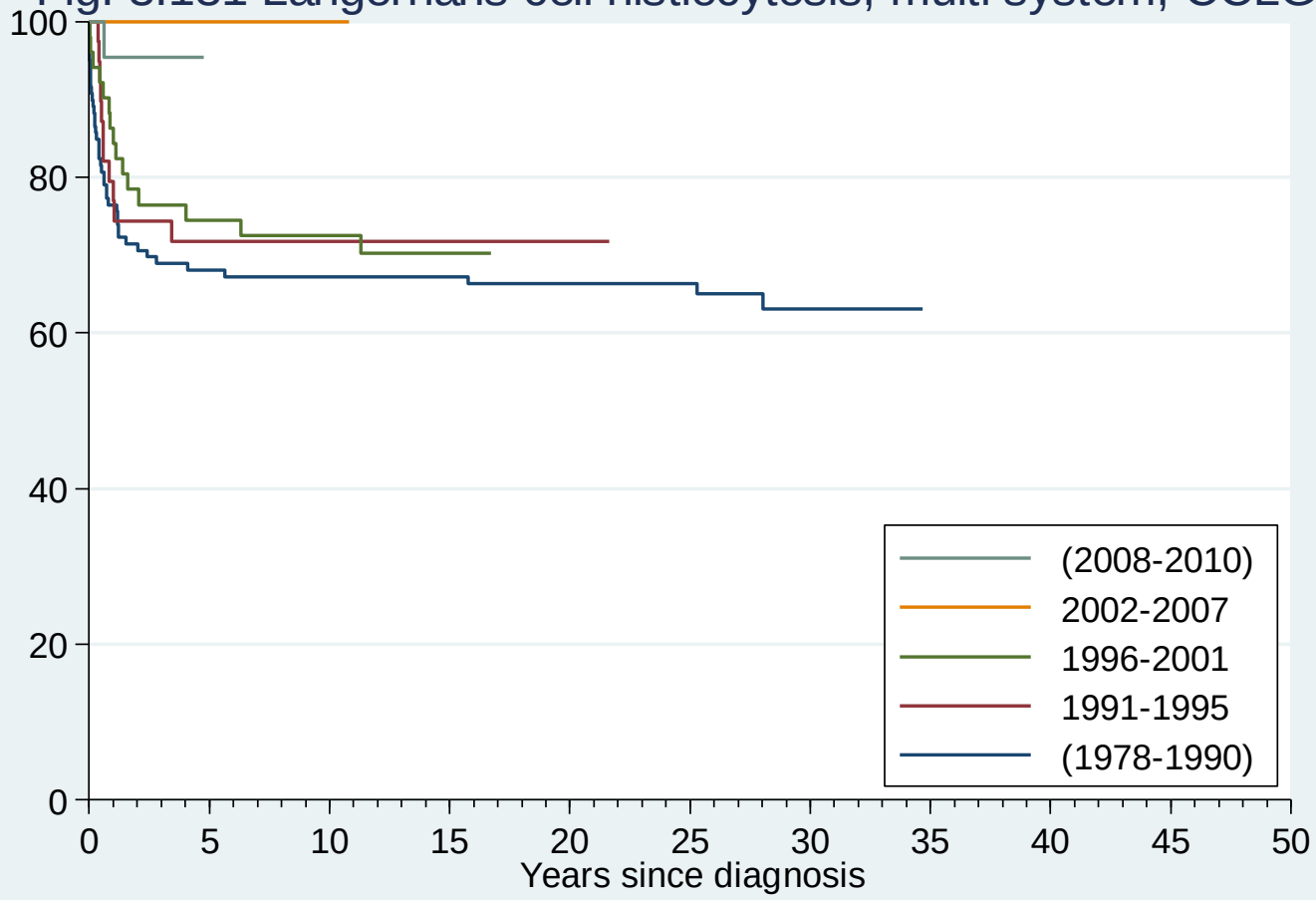


Fig. 3.131 Langerhans cell histiocytosis, multi system, CCLG



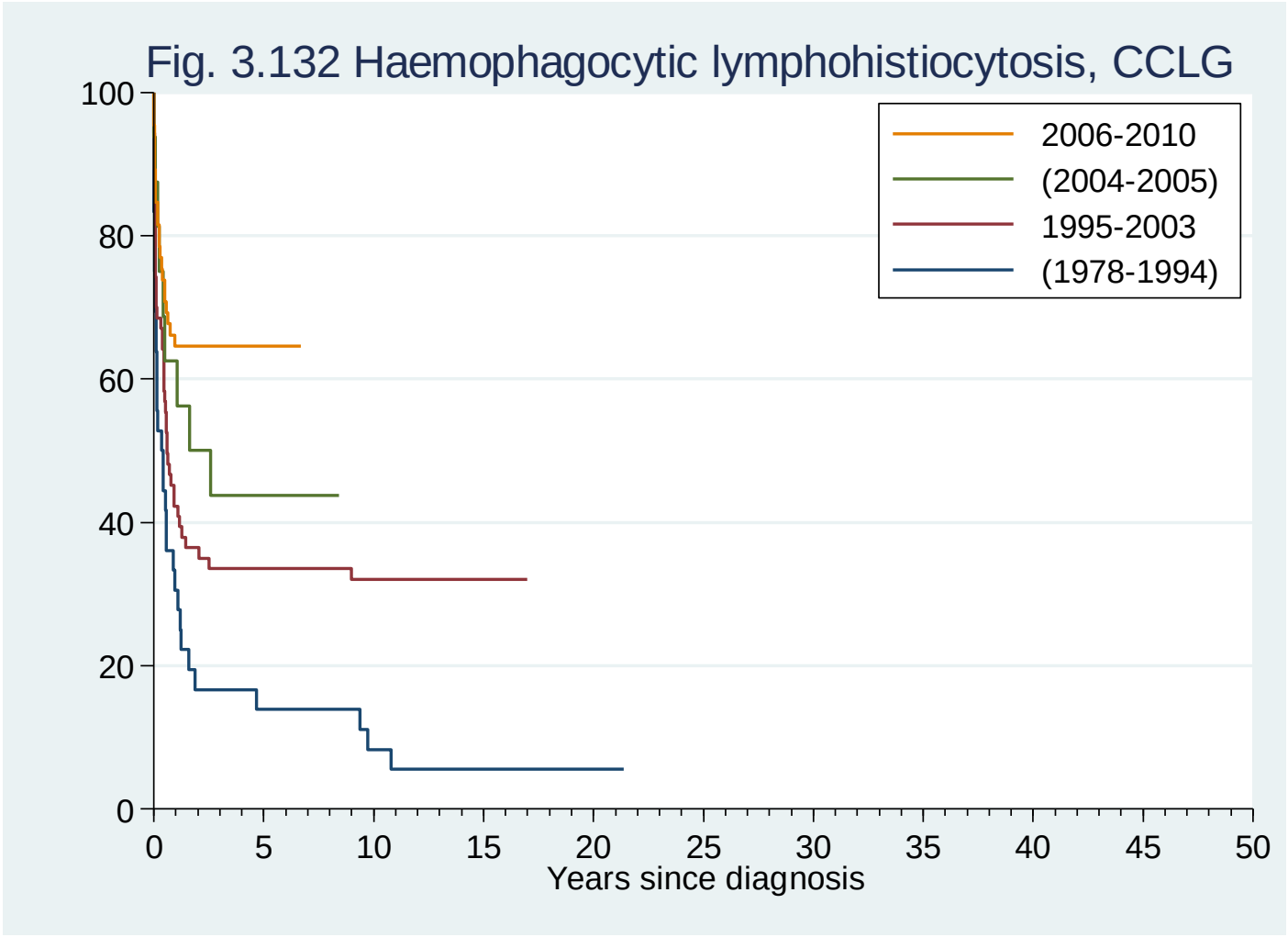


Fig. 3.132 Haemophagocytic lymphohistiocytosis, CCLG
100
80
60
40
20
0
0
5
10
15
20
25
30
35
40
45
50
Years since diagnosis
2006-2010
(2004-2005)
1995-2003
(1978-1994)

4. Publications

The NRCT data have been used in a wide range of publications. Those published in 2010 onwards, together with other publications by NRCT staff, are listed below.

(i) Journal articles

Anoop P, Sankpal S, Stiller C, Tewari S, Lancaster DL, Khabra K, Taj M (2012) Outcome of childhood relapsed or refractory mature B-cell non-Hodgkin lymphoma and acute lymphoblastic leukaemia. *Leuk Lymphoma* **53** (10): 1882-1888

Brennan B, Stevens M, Kelsey A, Stiller CA (2010) Synovial sarcoma in childhood and adolescence: A retrospective series of 77 patients registered by the Children's Cancer and Leukaemia Group between 1991 and 2006. *Pediatr Blood Cancer* **55** (1): 85-90

Breslow NE, Lange JM, Friedman DL, Green DM, Hawkins MM, Murphy MFG, Neglia JP, Olsen JH, Peterson SM, Stiller CA, Robison LL (2010) Secondary malignant neoplasms following Wilms Tumor: an International Collaborative Study. *Int J Cancer* **127** (3): 657-666

Chumas P, Kenny T, Stiller C (2011) Subspecialisation in neurosurgery-does size matter? *Acta Neurochir (Wien)* **153** (6): 1231-1236

Crocetti E, Trama A, Stiller C, Caldarella A, Soffietti R, Jaal J, Weber DC, Ricardi U, Slowinski J, Brandes A, RARECARE working group (2012) Epidemiology of glial and non-glial brain tumours in Europe. *Eur J Cancer* **48** (10): 1532-1542

England RJ, Haider N, Vujanic GM, Kelsey A, Stiller CA, Pritchard-Jones K, Powis M (2011) Mesoblastic Nephroma: A Report of the United Kingdom Children's Cancer and Leukaemia Group (CCLG). *Pediatr Blood Cancer* **56** (5): 744-748

Frobisher C, Lancashire ER, Reulen RC, Winter DL, Stevens MC, Hawkins MM, on behalf of the British Childhood Cancer Survivor Study (2010) Extent of alcohol consumption among adult survivors of childhood cancer: the British Childhood Cancer Survivor Study. *Cancer Epidemiol Biomarkers Prev* **19** (5): 1174-1184

Frobisher C, Gurung PM, Leiper A, Reulen RC, Winter DL, Taylor AJ, Lancashire ER, Woodhouse CR, Hawkins MM (2010) Risk of bladder tumours after childhood cancer: the British Childhood Cancer Survivor Study. *BJU Int* **106** (7): 1060-1069

Gatta G, Ferrari A, Stiller CA, Pastore G, Bisogno G, Trama A, Capocaccia R, The RARECARE Working Group (2012) Embryonal cancers in Europe. *Eur J Cancer* **48** (10): 1425-1433

Jones EA, Stewart A, Stiller C, Douglas F, Bown N (2011) Wilms tumor incidence in children with 2q terminal deletions: A cohort study. *Am J Med Genet Part A* **155A** (9): 2221-2223

Keegan TJ, Bunch KJ, Vincent TJ, King JC, O'Neill KA, Kendall GM, MacCarthy A, Fear NT, Murphy MFG (2012) Case-control study of paternal occupation and risk of childhood leukaemia in Great Britain, 1962-2006. *Br J Cancer* **107** (9): 1652-1659

Kendall GM, Little MP, Wakeford R, Bunch KJ, Miles JCH, Vincent TJ, Meara JR, Murphy MFG (2013) A record-based case-control study of natural background radiation and the incidence of childhood leukaemia and other cancers in Great Britain during 1980-2006. *Leukemia* **27** (1): 3-9

Kendall GM, Little MP, Wakeford R (2011) Numbers and proportions of leukemias in young people and adults induced by radiation of natural origin. *Leuk Res* **35** (8): 1039-1043

Kheifets L, Ahlbom A, Crespi CM, Feychting M, Johansen C, Monroe J, Murphy MF, Oksuzyan S, Preston-Martin S, Roman E, Saito T, Savitz D, Schüz J, Simpson J, Swanson J, Tynes T, Verkasalo P, Mezei G (2010) A Pooled Analysis of Extremely Low-Frequency Magnetic Fields and Childhood Brain Tumors. *Am J Epidemiol* **172** (7): 752-761

Kheifets L, Ahlbom A, Crespi CM, Draper G, Hagihara J, Lowenthal RM, Mezei G, Oksuzyan S, Schüz J, Swanson J, Tittarelli A, Vinceti M, Wunsch Filho V (2010) Pooled analysis of recent studies on magnetic fields and childhood leukaemia. *Br J Cancer* **103** (7): 1128-1135

Kroll ME, Swanson J, Vincent TJ, Draper GJ (2010) Childhood cancer and magnetic fields from high-voltage power lines in England and Wales: a case-control study. *Br J Cancer* **103** (7): 1122-1127

Kroll ME, Murphy MFG, Carpenter LM, Stiller CA (2011) Childhood cancer registration in Britain: capture-recapture estimates of completeness of ascertainment. *Br J Cancer* **104** (7): 1227-1233

Kroll ME, Stiller CA, Murphy MFG, Carpenter LM (2011) Childhood leukaemia and socioeconomic status in England and Wales 1976-2005: evidence of higher incidence in relatively affluent communities persists over time. *Br J Cancer* **105** (11): 1783-1787

Kroll ME, Carpenter LM, Murphy MFG, Stiller CA (2012) Effects of changes in diagnosis and registration on time trends in recorded childhood cancer incidence in Great Britain. *Br J Cancer* **107** (7): 1159-1162

Kroll ME, Stiller CA, Richards S, Mitchell C, Carpenter LM (2012) Evidence for under-diagnosis of childhood acute lymphoblastic leukaemia in poorer communities within Great Britain. *Br J Cancer* **106** (9): 1556-1559

Lancashire ER, Frobisher C, Reulen RC, Winter DL, Glaser A, Hawkins MM (2010) Educational attainment among adult survivors of childhood cancer in great britain: a population-based cohort study. *J Natl Cancer Inst* **102** (4): 254-270

Little MP, Kleinerman RA, Stiller CA, Li G, Kroll ME, Murphy MFG (2012) Analysis of retinoblastoma age incidence data using a fully stochastic cancer model. *Int J Cancer* **130** (3): 631-640

MacCarthy A, Bunch KJ, Fear NT, King JC, Vincent TJ, Murphy MFG (2010) Paternal occupation and neuroblastoma: a case-control study based on cancer registry data for Great Britain 1962-1999. *Br J Cancer* **102** (3): 615-619

McNally RJQ, Blakey K, Parslow RC, James PW, Goméz Pozo B, Stiller C, Vincent TJ, Norman P, McKinney PA, Murphy MF, Craft AW, Feltbower RG (2012) Small-area analyses of bone cancer diagnosed in Great Britain provide clues to aetiology. *BMC cancer* **12** 270

O'Neill KA, Bunch KJ, Vincent TJ, Spector LG, Moorman AV, Murphy MFG (2012) Immunophenotype and cytogenetic characteristics in the relationship between birth weight and childhood leukemia. *Pediatr Blood Cancer* **58** (1): 7-11

Rebholz CE, Reulen RC, Toogood AA, Frobisher C, Lancashire ER, Winter DL, Kuehni CE, Hawkins MM (2011) Health care use of long-term survivors of childhood cancer: the british childhood cancer survivor study. *J Clin Oncol* **29** (31): 4181-4188

Reulen RC, Winter DL, Frobisher C, Lancashire ER, Stiller CA, Jenney ME, Skinner R, Stevens MC, Hawkins MM, for the British Childhood Cancer Survivor Study Steering Group (2010) Long-term cause-specific mortality among survivors of childhood cancer. *JAMA* **304** (2): 172-179

Reulen RC, Frobisher C, Winter DL, Kelly J, Lancashire ER, Stiller CA, Pritchard-Jones K, Jenkinson HC, Hawkins MM, for the British Childhood Cancer Survivor Study Steering Group (2011) Long-term risks of subsequent primary neoplasms among survivors of childhood cancer. *JAMA* **305** (22): 2311-2319

Satgé D, Stiller CA, Rutkowski S, von Bueren AO, Lacour B, Sommelet D, Nishi M, Massimino M, Garré ML, Moreno F, Hasle H, Jakab Z, Greenberg M, von der Weid N, Kuehni C, Zurriaga O, Vicente ML, Peris-Bonet R, Benesch M, Vekemans M, Sullivan SG, Rickert C (2013) A very rare cancer in Down syndrome: medulloblastoma. Epidemiological data from 13 countries. *J Neurooncol* **112** (1): 107-114

Scott RH, Murray A, Baskcomb L, Turnbull C, Loveday C, Al-Saadi R, Williams R, Breatnach F, Gerrard M, Hale J, Kohler J, Lapunzina P, Levitt GA, Picton S, Pizer B, Ronghe MD, Traunecker H, Williams D, Kelsey A, Vujanic GM, Sebire NJ, Grundy P, Stiller CA, Pritchard Jones K, Douglas J, Rahman N (2012) Stratification of Wilms tumor by genetic and epigenetic analysis. *Oncotarget* **3** (3): 327-335

Shah A, Stiller C, Lancaster D, Vincent T, Coleman MP (2010) Leukaemia survival trends in children with Down's syndrome in Great Britain, 1971-2000: a population-based study. *J Epidemiol Community Health* **64** (7): 604-609

Shah A, Diggens N, Stiller C, Murphy D, Passmore SJ, Murphy MFG (2011) Place of death and hospital care for children who died of cancer in England, 1999-2006. *Eur J Cancer* **47** (14): 2175-2181

Slade I, Stephens P, Douglas J, Barker K, Stebbings L, Abbaszadeh F, Pritchard-Jones K, FACT collaboration., Cole R, Pizer B, Stiller C, Vujanic G, Scott RH, Stratton MR, Rahman N (2010) Constitutional translocation breakpoint mapping by genome-wide paired-end sequencing identifies HACE1 as a putative Wilms tumor susceptibility gene. *J Med Genet* **47** (5): 342-347

Slade I, Bacchelli C, Davies H, Murray A, Abbaszadeh F, Hanks S, Barfoot R, Burke A, Chisholm J, Hewitt M, Jenkinson H, King D, Morland B, Pizer B, Prescott K, Sagger A, Side L, Traunecker H, Vaidya S, Ward P, Futreal PA, Vujanic G, Nicholson AG, Sebire N, Turnbull C, Priest JR, Pritchard-Jones K, Houlston R, Stiller C, Stratton MR, Douglas J, Rahman N (2011) DICER1 syndrome: clarifying the diagnosis, clinical features and management implications of a pleiotropic tumour predisposition syndrome. *J Med Genet* **48** (4): 273-278

Slade I, Murray A, Hanks S, Kumar A, Walker L, Hargrave D, Douglas J, Stiller C, Izatt L, Rahman N (2011) Heterogeneity of familial medulloblastoma and contribution of germline PTCH1 and SUFU mutations to sporadic medulloblastoma. *Fam Cancer* **10** (2): 337-342

Stiller CA, Trama A, Serraino D, Rossi S, Navarro C, Chirlaque MD, Casali PG, The RARECARE Working Group (2013) Descriptive Epidemiology of Sarcomas in Europe. Report from the RARECARE Project. *Eur J Cancer* **49** (3): 684-695

Stiller CA, Kroll ME, Pritchard-Jones K (2012) Population survival from childhood cancer in Britain during 1978–2005 by eras of entry to clinical trials. *Ann Oncol* **23** (9): 2464-2469

Taylor AJ, Little MP, Winter DL, Sugden E, Ellison DW, Stiller CA, Stovall M, Frobisher C, Lancashire ER, Reulen RC, Hawkins MM (2010) Population-based risks of CNS tumors in survivors of childhood cancer: The British Childhood Cancer Survivor Study. *J Clin Oncol* **28** (36): 5287-5293

Turnbull C, Perdeaux ER, Pernet D, Naranjo A, Renwick A, Seal S, Munoz-Xicola RM, Hanks S, Slade I, Zachariou A, Warren-Perry M, Ruark E, Gerrard M, Hale J, Hewitt M, Kohler J, Lane S, Levitt G, Madi M, Morland B, Neefjes V, Nicholson J, Picton S, Pizer B, Ronghe M, Stevens M, Traunecker H, Stiller CA, Pritchard-Jones K, Dome J, Grundy P, Rahman N (2012) A genome-wide association study identifies susceptibility loci for Wilms tumor. *Nat Genet* **44** (6): 681-684

Visser O, Trama A, Maynadié M, Stiller C, Marcos-Gragera R, De Angelis R, Mallone S, Tereanu C, Allemani C, Ricardi U, Schouten HC, The RARECARE Working Group (2012) Incidence, survival and prevalence of myeloid malignancies in Europe. *Eur J Cancer* **48** (17): 3257-3266

Wakeford R, Darby SC, Murphy MFG (2010) Temporal trends in childhood leukaemia incidence following exposure to radioactive fallout from atmospheric nuclear weapons testing. *Radiat Environ Biophys* **49** (2): 213-247

(ii) Book chapters etc

AGIR. Risk of Solid Cancers following Radiation Exposure: Estimates for the UK Population. Report of the independent Advisory Group on Ionising Radiation. RCE-19, 1-258. 2011. Chilton, HPA.

Brennan B, Stiller C (2010) Rare tumours. In *Pediatric Hematology and Oncology. Scientific Principles and Clinical Practice*, Estlin EJ, Gilbertson RJ, Wynn RF (eds) pp 319-332. Wiley-Blackwell: Chichester.

Stiller CA, Shah A (2012) The epidemiology of cancer in children and adolescents. In *Cancer in children: clinical management, 6th edition*, Stevens MCG, Caron HN, Biondi A (eds) pp 1-13. Oxford University Press: Oxford.

(iii) NCIN Reports and Data Briefings (all available on NCIN website)

Reports

Shared care and survival from childhood cancer in the UK 1997-2009	2013
Multiple neoplasms in patients with cancer diagnosed during childhood	2012
Supra-regional referral patterns of childhood cancer patients	2012

Data briefings

Short-term survival of children with cancer	2013
Occurrence of cancer among five-year survivors of childhood cancer	2012
Place of death for children, teenagers and young adults with cancer in England	2011
Survival of children, teenagers and young adults with cancer in England	2011

Authorship and Acknowledgements

This report was written by Charles Stiller, many of the analyses and tabulations were done by Nicole Diggens and Tim Vincent, and data collection was managed by Anita Bayne, all at the Childhood Cancer Research Group (CCRG). We are, as always, very grateful to all those organisations and individuals that have provided information on which the report is based.