National Cancer Registry Ireland **Cancer Trends**

Primary Bone Cancer

Case numbers, tumour sites and histological types

Between 1994 and 2012, 671 cases of invasive primary bone cancer were diagnosed in Ireland, an annual average of 15 female and 21 male cases (Table 1). Incidence rate in males was almost 50% higher than in females and lifetime risk was approximately 1 in 2,000 for females and 1 in 1,300 for males. At the end of 2012, there were 348 people alive in the Irish population that had had a diagnosis of primary bone cancer at some time since 1994.

Table 1. Annual average number and incidence rate of all primary invasive bone cancers in Ireland, 1994-2012

	females	males	total
cases per year	15	21	35
incidence rate	0.7	1.0	0.9
cumulative lifetime risk (1 in -)	1988	1262	1543
% of all cancers	0.2%	0.3%	0.2%
19 year prevalence at end 2012	145	203	348

Over half of all bone cancers were located in the limbs, 40% in the legs and 12% in the arms and shoulders. A further 16% were diagnosed in the pelvis and lower bones of the vertebral column (Table 2). The bulk of tumours were osteosarcoma, chondrosarcoma or Ewing's sarcoma subtypes. Over 90% of all cases were microscopically confirmed and 9% were unspecified. Little difference was observed between males and females.

Table 2. Total numbers of primary bone cancers by tumour site and histological subtype, 1994-2012

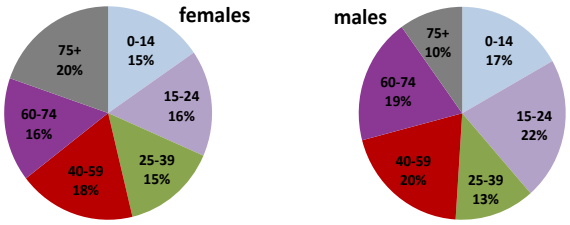
	females	males	total	% all
Body site				
upper limbs	28	55	83	12%
lower limbs	109	157	266	40%
skull & face	20	26	46	7%
mandible	8	20	28	4%
vertebral column	19	25	44	7%
ribs, sternum & clavicle	19	18	37	6%
pelvis, sacrum & coccyx	46	65	111	16%
unspecified	30	26	56	8%
Histological group				
sarcoma				
osteosarcoma	81	118	199	30%
chondrosarcoma	82	105	187	28%
Ewing's sarcoma	41	81	122	18%
giant cell	7	6	13	2%
other	15	17	32	5%
unspecified	8	9	17	2%
*other (non-sarcoma)	15	26	41	6%
unspecified	30	30	60	9%
All invasive	279	392	671	

** includes chordoma, odontogenic tumours, ameloblastoma*

Age profile

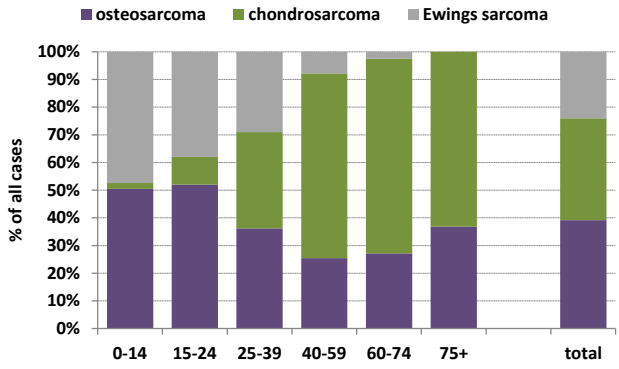
Bone cancers have a younger age profile than many other cancer types: 15% of female and 17% of male patients were aged under 15 years at diagnosis (Figure 1). Approximately half of all patients were diagnosed before age 40. Although the age distribution was broadly similar between males and females, there was a slightly greater proportion of elderly (aged ≥75) female patients compared to males.

Figure 1. Age distribution of bone cancer patients, 2007-2011



The majority of tumours in children and young adults consisted of osteosarcoma and Ewings sarcoma, consistent with information published in the literature[1] (Figure 2). Chondrosarcomas were more common in patients over 40 years and Ewing's sarcomas were rarely diagnosed in patients over 60.

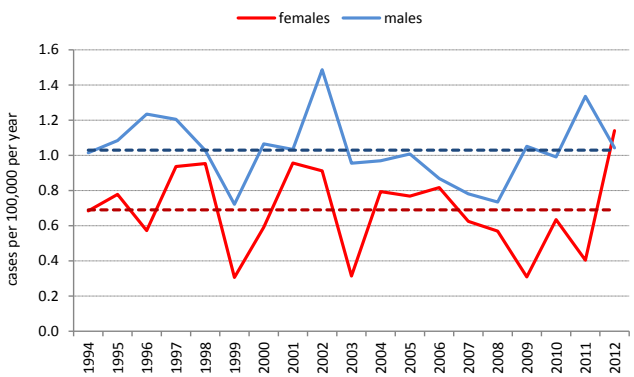
Figure 2. Percentage composition of the 3 main subtypes by age group, 1994–2012



Time trends in incidence and mortality

There has been considerable variation in incidence rates for both sexes since 1994 with no clear overall trend evident (Figure 3). Incidence in males has been higher than in females each year but wide fluctuations were observed in both sexes. There was also fairly little change in the percentage composition of subtypes or tumour sites over time.

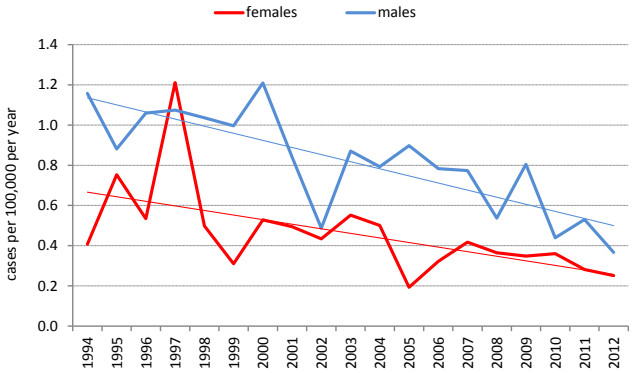
Figure 3. Trends in bone cancer incidence, 1994–2012



Dashed lines show average 1994-2012 rate

Despite the lack of any real change in incidence, there has been a marked decline in mortality rates over time (Figure 4). From 1994 to 2012 mortality rates declined by 4.0% per year in females and 4.5% per year in males. Total numbers of deaths have declined from an average of 30 per year in the late 1990's to approximately 20 deaths per year between 2008 and 2012 inclusive. As a result mortality/incidence ratios have fallen from 0.9 in both sexes during the 1990's to 0.7 (males) and 0.5 (females) in 2008-2012.

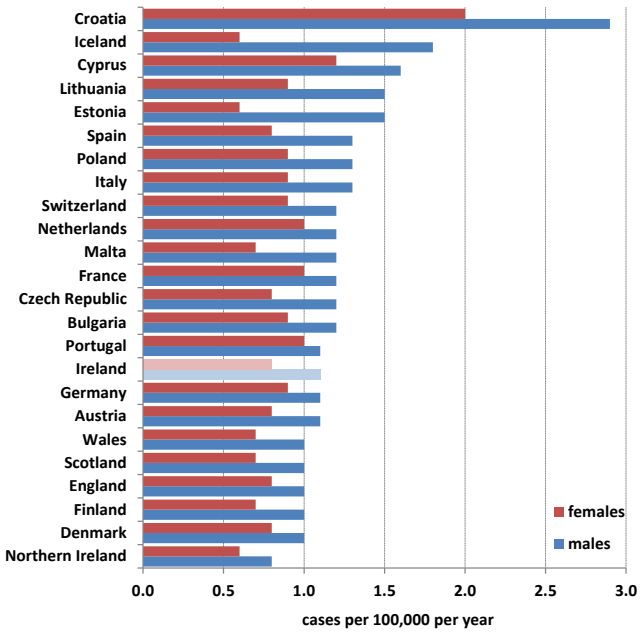
Figure 4. Trends in mortality from primary bone cancer, 1994–2012



International variation in incidence

Comparing incidence data for the period 1998-2006 across Europe[2], there was little variation in males with most countries ranging from 0.8 to 1.2 cases per 100,000 per year (Figure 5). However male rates in Croatia were over 160% higher than the European average of 1.1. Similarly incidence rates for females in Croatia were 150% higher than the European average (0.8 cases per 100,000 per year). Ireland ranked close to the European average for both males and females. Incidence in males was higher than in females in all countries although male : female ratios varied widely, being lowest in Portugal, Germany, France and Netherlands and highest in Iceland, Estonia, Lithuania and Malta. The considerably higher incidence rates in Croatia and variability in the male : female ratios between countries may relate to some inconsistencies in registration practices.

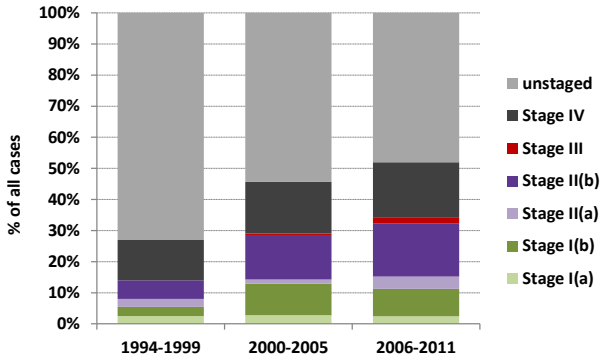
Figure 5. Incidence of bone cancer in Europe, 1998-2006[2]



Stage at diagnosis

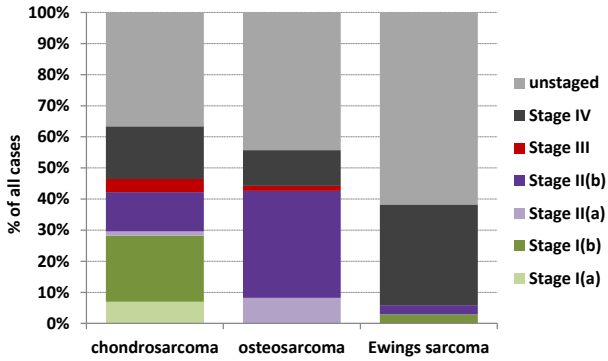
A large number of bone cancers are not staged; in the 1990's almost 3 in every 4 cases were of unknown stage. Although the proportion of these has decreased over time, in 2006-2011 almost half of all cases still remained unstaged (Figure 6). In 2006-2011 just over 10% of all diagnosed cases were low grade (stage I) tumours and 20% were stage II (high grade). Only 2% of all cases were classed as stage III and 18% already had regional or distant metastases at diagnosis (stage IV). Males and females had very similar stage distribution patterns.

Figure 6. Variation in the distribution of bone cancers by stage over time[3]



Tumour stage varied considerably by subtype and although over one-third of chondrosarcomas diagnosed between 2006 and 2011 remained unstaged, cancers of this subtype were generally diagnosed at an earlier stage than either osteosarcoma or Ewing's sarcoma (Figure 7). Two-thirds of Ewing's sarcomas were unstaged and of the cases that were staged, almost all had regional or distant metastasis.

Figure 7. Percentage of bone cancer by stage for the main subtypes, 2006-2011[3]



Treatment

All but 12% of patients diagnosed between 2006 and 2011 had tumour-directed surgery, chemo- or radio-therapy (Table 3). Surgery is the principal treatment for bone cancer, and over two-thirds of all patients had surgical removal of their tumour. Numbers having surgery varied by patient age and subtype. Patients diagnosed with Ewing's sarcoma (most common in people aged under 25) were least likely to have surgery, with just 53% treated.

Almost half of all patients underwent chemotherapy. As chondrosarcoma does not normally respond well to chemotherapy[4], only 14% of these patients were treated. Chondrosarcoma is more common in older patients and this may contribute to the lower percentage of older patients undergoing chemotherapy. All 34 patients diagnosed with Ewing's sarcoma during this period had chemotherapy.

Overall one-third of patients had radiotherapy, although this varied from 20% of chondrosarcoma to 74% of Ewing's sarcoma patients. There was less variation between age groups than was observed for surgery and chemotherapy.

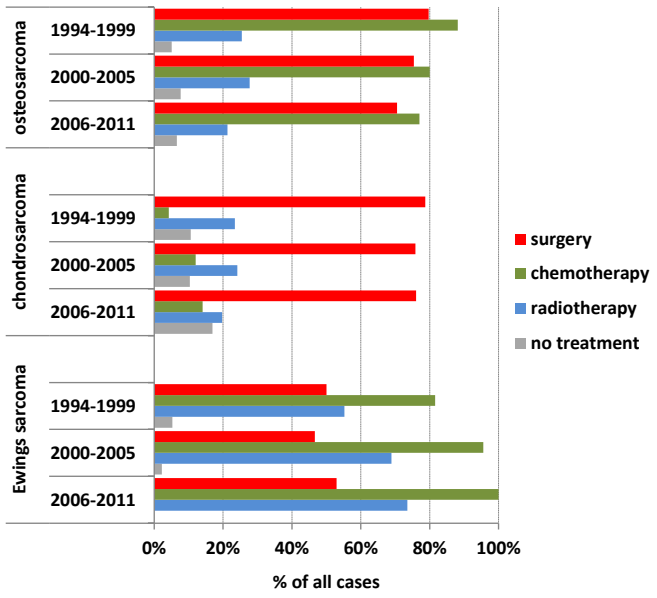
All patients with Ewing's sarcoma diagnosed during this period had at least some form of tumour directed therapy. However 17% of chondrosarcoma patients remained untreated. Their generally older age profile is also reflected in the increasing proportion of untreated patients with age.

Table 3. Variation in treatment by patient age and subtype, 2006-2011[3]

	surgery	chemo therapy	radio therapy	none
All cases	67%	48%	33%	12%
<25 years	67%	90%	33%	0%
25-39 years	74%	62%	38%	5%
40-59 years	80%	32%	32%	10%
60-74 years	62%	28%	31%	21%
75+ years	39%	0%	32%	39%
osteosarcoma	70%	77%	21%	7%
chondrosarcoma	76%	14%	20%	17%
Ewing's sarcoma	53%	100%	74%	0%

There was a 12.5% decline in the proportion of osteosarcoma patients undergoing surgery and chemotherapy over time but there was little change in the treatments received by patients with chondrosarcoma (Figure 8). The greatest change in treatment over time was observed for Ewing's sarcoma where chemotherapy increased from 82% to 100% between 1994-1999 and 2006-2011. Radiotherapy also increased over time for these patients, from 55% to 74%.

Figure 8. Variation in treatment over time for patients diagnosed with osteosarcoma, chondrosarcoma and Ewing's sarcoma

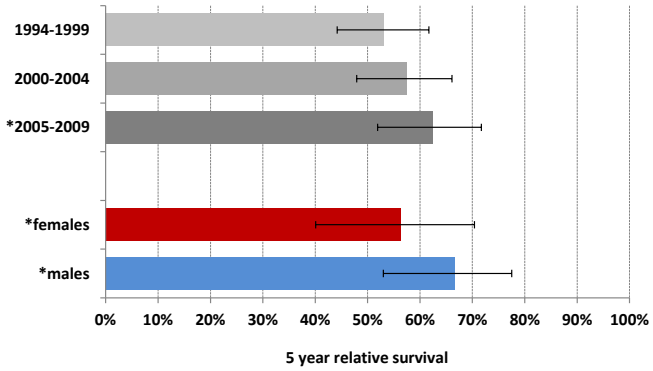


Survival[5]

Survival rates for all primary bone cancers have improved over time and most recent estimates indicate a 5 year relative survival of 62.5% (Figure 9). During the most recent period for which data is available (2005-2009), survival in males was higher than in females, although this was not statistically significant.

Recently published figures indicate a European average 5 year survival for primary bone cancer of 53% (age standardised for 2000-2007)[6]. Similarly age standardised survival rates in Ireland indicate values of 50.5% for the period 2000-2004 and 59.0% for 2005-2009.

Figure 9. Five year relative survival (with 95% confidence intervals) for all primary bone cancer[5]



**females and males for 2005-2009 period only*
rates shown are all unstandardized

References and notes

1. Miller RW et al. 1996 Bone cancer. In: Schottenfeld D, Fraumeni JF, editors. *Cancer Epidemiology and Prevention*. 2nd ed. New York: Oxford University Press
2. EUREG database, European Cancer Observatory (ECO), International Agency for Research on Cancer. http://eco.iarc.fr/EUREG/Default.aspx
3. Complete data on stage and treatment available to 2011 only
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5. All patients followed up to end 2011
6. DeAngelis, R et al 2014. Cancer survival in Europe 1999–2007 by country and age: results of EUROCARE-5—a population-based study. Lancet Oncology 15 (1): 23-34

