4 Cancer in children

The German Childhood Cancer Registry (DKKR) has been based at the Institute of Medical Biostatistics, Epidemiology and Informatics at the Johannes Gutenberg University, Mainz, since beginning its work in 1980. Close cooperation with the Society for Paediatric Oncology and Haematology (GPOH) and the associated hospitals was already part of the DKKR’s original conception. This is a characteristic feature of the registry which cannot be easily applied to adult oncology. A nationwide, population-based cancer registry with a high level of data quality and a degree of completeness of over 95% has been built up, covering the entire Federal Republic. The DKKR thus meets international standards for population-based cancer registries. Tumours of the central nervous system (CNS tumours) are an exception here; completeness in this field is slightly lower. A further characteristic of the DKKR is that it has implemented an active, open-end, long-term follow-up system which continues long into adulthood. In this way the registry also provides the basis for research into long-term effects, secondary tumours, and for studies with long-term survivors in general.

The registry population comprises children who are diagnosed with a malignant disease or a histologically benign brain tumour before their 15th birthday and are part of the resident population of the Federal Republic of Germany when diagnosed. Cancer cases in eastern Germany have been included since 1991 in cooperation with the joint cancer registry (GKR) of Berlin, Brandenburg, Mecklenburg-Western Pomerania, Saxony-Anhalt, Saxony and Thuringia. The current data basis consists of over 41,000 cancer cases.

Incidence of childhood cancers in Germany

About 1,800 cases are newly diagnosed every year in Germany. With an overall population of approx. 13 million children under the age of 15, this means an annual incidence of about 14 per 100,000 children in this age group. The likelihood that a newborn child will develop a malignant disease within the first 15 years of its life is 224:100,000 (0.2%). In other words, a malignant cancer disease is diagnosed in approx. one in 500 children up to their 15th birthday.

Range of diagnoses

The pattern of cancer diagnoses in children is completely different from that of adults. For example, children are mostly affected by embryonal tumours (neuroblastomas, retinoblastomas, nephroblastomas, medulloblastomas, embryonic rhabdomyosarcomas or germ-cell tumours); carcinomas, by contrast, are very rare in childhood (less than 2% of all malignant diseases). The largest diagnostic groups are leukaemias (34.1%), CNS tumours (22.1%) and lymphomas (11.8%).

Figure 4.1
Cancer in children
Determined for the period 1997–2006

Germ-cell tumours 3.2%
Bone tumours 4.4%
Other Diagnoses 4.7%
Kidney tumours 5.6%
Soft tissue (sarkome) 6.2%
Peripheral neural cell tumours 7.8%
Lymphomas 11.8%
CNS tumours 22.1%
Leukaemias

Leukaemias make up more than a third of all cancers among the under-15s. The most common single diagnosis overall is acute lymphatic leukaemia (ALL, 27.0%). It occurs more than twice as frequently among children under the age of four as in the other age groups. 4.8% of all childhood malignancies are acute myeloid leukaemias (AML). AML is most common...
Figure 4.4
Trends in the incidence of selected diagnostic groups and for all childhood malignancies
Number of cases per 100,000 (age-standardized), including eastern Germany since 1991

Figure 4.5
Trends in the incidence of childhood leukaemias
Number of cases per 100,000 (age-standardized), including eastern Germany since 1991

in children under the age of two. The survival rates for AML are markedly lower than for ALL. The causes of leukaemias in childhood remain largely uncertain, even today. For a long time environmental influences were suspected of causing childhood leukaemias. In the meantime it has been shown that the number of cases caused by most environmental factors (low-dose ionizing radiation, non-ionizing radiation and pesticides) is quite small after all, even if an association with leukaemias in childhood cannot be ruled out. A number of clues have in the meantime strengthened hypotheses that assign a central role to infectious pathogens in the development of leukaemias in childhood. Especially children with an insufficiently modulated immune system in babyhood can have a higher risk of developing leukaemia.


**CNS tumours**

The most common single diagnoses among CNS tumours are astrocytomas (total: 10.2%), intracranial and intraspinal embryonal tumours (5.0%) and ependymomas (2.2%). The increase in the incidence of CNS tumours observed in a number of western countries over the past decades may be connected with general changes in environmental factors and related exposure. For example, a number of epidemiological studies are looking into the possible influence of ionizing radiation, electromagnetic fields, pesticides, the mother’s diet and genetic aspects.

**Lymphomas**

The most common lymphomas are non-Hodgkin lymphomas (NHL, total: 5.5%) and Hodgkin’s disease (4.9%). The chances of survival with Hodgkin's disease are among the best in paediatric oncology. Children with congenital or acquired immunodeficiency and those who have had immunosuppressive therapy are at increased risk of developing NHL. An association is suspected between lymphomas and ionizing radiation; this has not, however, been substantiated.

**Other common malignant diseases**

Other common malignant diseases in childhood include neuroblastomas (nerve-cell tumours), Wilms’ tumours or nephroblastomas (kidney tumours), germ-cell tumours, bone tumours and rhabdomyosarcomas (tumours of the skeletal musculature). Among these malignancies, the prognosis for children with nephroblastoma or a germ-cell tumour is much more favourable than for the others.

**Survival**

Children with cancer make up fewer than 1% of all cancer patients. However, malignant neoplasms are the second most common cause of death among children. Fortunately, the survival rates have improved dramatically over the last 30 years thanks to significantly more differentiated diagnostics and the use of multimodal therapy.

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**Table 4.1**

*Number of cancer cases*

*Percentage incidence and survival rates, determined for the period 1997–2006*

<table>
<thead>
<tr>
<th>Cancer sites</th>
<th>Incidence*</th>
<th>Survival rate in percent after 3 years</th>
<th>Survival rate in percent after 5 years</th>
<th>Survival rate in percent after 10 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hodgkin lymphomas</td>
<td>0.7</td>
<td>97</td>
<td>97</td>
<td>95</td>
</tr>
<tr>
<td>Germ-cell tumours</td>
<td>0.5</td>
<td>95</td>
<td>94</td>
<td>92</td>
</tr>
<tr>
<td>Lymphatic leukaemias</td>
<td>4.1</td>
<td>91</td>
<td>88</td>
<td>85</td>
</tr>
<tr>
<td>Nephroblastomas</td>
<td>0.9</td>
<td>91</td>
<td>90</td>
<td>89</td>
</tr>
<tr>
<td>Non-Hodgkin lymphomas</td>
<td>0.8</td>
<td>88</td>
<td>87</td>
<td>86</td>
</tr>
<tr>
<td>Neuroblastomas and ganglioneuroblastomas</td>
<td>1.3</td>
<td>83</td>
<td>78</td>
<td>75</td>
</tr>
<tr>
<td>Osteosarcomas</td>
<td>0.3</td>
<td>82</td>
<td>73</td>
<td>68</td>
</tr>
<tr>
<td>Astrocytomas</td>
<td>1.5</td>
<td>79</td>
<td>77</td>
<td>73</td>
</tr>
<tr>
<td>Rhabdomyosarcomas</td>
<td>0.5</td>
<td>78</td>
<td>74</td>
<td>71</td>
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<tr>
<td>Ewing’s tumours &amp; related bone sarcomas</td>
<td>0.3</td>
<td>74</td>
<td>69</td>
<td>65</td>
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<tr>
<td>Intracranial &amp; intraspinal embryonal tumours</td>
<td>0.8</td>
<td>68</td>
<td>61</td>
<td>52</td>
</tr>
<tr>
<td>Acute myeloid leukaemias</td>
<td>0.7</td>
<td>65</td>
<td>62</td>
<td>60</td>
</tr>
<tr>
<td>All malignancies</td>
<td>15.0</td>
<td>84</td>
<td>81</td>
<td>77</td>
</tr>
</tbody>
</table>

* related to 100,000 children under the age of 15, age-standardized to the west German population in 1987
In the early 1980s the chances of children with cancer still being alive five years after diagnosis were 67%; this figure has risen to 80% in the meantime. Looking at all patients of the registry population who were diagnosed between 1997 and 2006 and followed up, the overall chance of survival is 81% after five years, 77% after ten years, and 76% after 15 years.

The encouraging increase in the number of long-term survivors is increasingly focusing attention on the long-term observation of former paediatric cancer patients. The DKKR provides an ideal data base for carrying out studies with long-term survivors. As the above figures show, it is already possible to provide information on long-term survival (after 5, 10 or 15 years) and to estimate the risk of developing a second malignancy after cancer in childhood. Examples of further research possibilities include the incidence of other long-term effects, such as the possible effects of therapy on fertility, and studies examining the health risks of the descendants of fathers and mothers who had cancer in childhood. About 12,000 of the more than 25,000 patients currently known to be alive have been under observation by the registry for at least twelve years. Over half of these patients are over 18 years old in the meantime, and are thus, in principle, available for studies with long-term survivors.

**Literature on childhood cancer**


