**Appendix:**

**Relationships describing bending trajectory of mortality from DNS**

If some maximal limit bounds congenital individual risks in the born population, then the following approximation may be valid according to the TCIR (Dolejs, 2001; Dolejs, 2003):

(7)

In the present study, *Di* decreased by three orders of magnitude, and *S*(*x*) was higher than 0.98 at the age of 10 years. Consequently, the changes in the denominator (number of living persons) with age can be neglected due to the changes in the numerator of equation (1). Based on the TCIR, we simultaneously assumed that the distribution of individual congenital risk *r* can be approximated by the relationship *f*(*r*) = *c*/*r*. Because the numerator is the partial derivative of *S*(*x*) with respect to the variable *x*, the variable *r* is only in the exponential term. (Variable *r* represents the denominator in the frequency function *f*(*r*), and it is also the result of the partial derivative in the numerator.) Furthermore, if the product *r*max.*x* is small in equation (7), then the theoretical mortality rate is approximately constant according to the following relationship (8) (Dolejs, 2001; Dolejs, 2003):

(8)

If a time unit of 1 year is used, and if r is represented per year, then the product rmax.x is unitless. The approximation (8) is valid if the product rmax.x is very small, relative to 1.

**Table 3**

**Congenital malformations of the nervous system according ICD10, Chapter XVII, codes: Q00-Q07**

**Q00 Anencephaly and similar malformations**

Q00.0 Anencephaly, Acephaly, Acrania, Amyelencephaly, Hemianencephaly, Hemicephaly,

Q00.1 Craniorachischisis

Q00.2 Iniencephaly

**Q01 Encephalocele,**

Incl.: encephalomyelocele, hydroencephalocele, hydromeningocele, cranial, meningocele, cerebral, meningoencephalocele, Excl.: Meckel-Gruber syndrome (Q61.9)

Q01.0 Frontal encephalocele

Q01.1 Nasofrontal encephalocele

Q01.2 Occipital encephalocele

Q01.8 Encephalocele of other sites

Q01.9 Encephalocele, unspecified

**Q02 Microcephaly**

Incl.: Hydromicrocephaly Micrencephalon Excl.: Meckel-Gruber syndrome (Q61.9)

**Q03 Congenital hydrocephalus**

Incl.:hydrocephalus in newborn Excl.: Arnold-Chiari syndrome (Q07.0), hydrocephalus: acquired NOS (G91.-), acquired, of newborn (P91.7) due to congenital toxoplasmosis (P37.1), with spina bifida (Q05.0-Q05.4)

Q03.0 Malformations of aqueduct of Sylvius, Aqueduct of Sylvius: anomaly, obstruction, congenital stenosis

Q03.1 Atresia of foramina of Magendie and Luschka, Dandy-Walker syndrome

Q03.8 Other congenital hydrocephalus

Q03.9 Congenital hydrocephalus, unspecified

**Q04 Other congenital malformations of brain**

Excl.: cyclopia (Q87.0), macrocephaly (Q75.3)

Q04.0 Congenital malformations of corpus callosum

Agenesis of corpus callosum

Q04.1 Arhinencephaly

Q04.2 Holoprosencephaly

Q04.3 Other reduction deformities of brain, Absence, Agenesis, Aplasia, Hypoplasia, Agyria, Hydranencephaly, Lissencephaly, Microgyria, Pachygyria, Excl.: congenital malformations of corpus callosum (Q04.0)

Q04.4 Septo-optic dysplasia

Q04.5 Megalencephaly

Q04.6 Congenital cerebral cysts, Porencephaly, Schizencephaly. Excl.: acquired porencephalic cyst (G93.0)

Q04.8 Other specified congenital malformations of brain, Macrogyria

Q04.9 Congenital malformation of brain, unspecified, Congenital: anomaly deformity disease or lesion, multiple anomalies

**Q05 Spina bifida**

Incl.: hydromeningocele (spinal), meningocele (spinal), meningomyelocele, myelocele, myelomeningocele, rachischisis, spina bifida (aperta)(cystica) , syringomyelocele, Excl.: Arnold-Chiari syndrome (Q07.0), spina bifida occulta (Q76.0)

Q05.0 Cervical spina bifida with hydrocephalus

Q05.1 Thoracic spina bifida with hydrocephalus, Spina bifida: dorsal, thoracolumbar

Q05.2 Lumbar spina bifida with hydrocephalus, Lumbosacral spina bifida with hydrocephalus

Q05.3 Sacral spina bifida with hydrocephalus

Q05.4 Unspecified spina bifida with hydrocephalus

Q05.5 Cervical spina bifida without hydrocephalus

Q05.6 Thoracic spina bifida without hydrocephalus, Spina bifida: dorsal NOS, thoracolumbar NOS

Q05.7 Lumbar spina bifida without hydrocephalus, Lumbosacral spina bifida NOS

Q05.8 Sacral spina bifida without hydrocephalus

Q05.9 Spina bifida, unspecified

**Q06 Other congenital malformations of spinal cord**

Q06.0 Amyelia

Q06.1 Hypoplasia and dysplasia of spinal cord, Atelomyelia, Myelatelia, Myelodysplasia of spinal cord

Q06.2 Diastematomyelia

Q06.3 Other congenital cauda equina malformations

Q06.4 Hydromyelia , Hydrorachis

Q06.8 Other specified congenital malformations of spinal cord

Q06.9 Congenital malformation of spinal cord, unspecified, Congenital: anomaly, deformity, disease or lesion

Q07 Other congenital malformations of nervous system,

Excl.:, familial dysautonomia [Riley-Day] (G90.1), neurofibromatosis (nonmalignant) (Q85.0)

Q07.0 Arnold-Chiari syndrome

Q07.8 Other specified congenital malformations of nervous system, Agenesis of nerve, Displacement of brachial plexus, Jaw-winking syndrome, Marcus Gunn syndrome

**Q07.9 Congenital malformation of nervous system, unspecified**

**Table 4**

**Diseases of the nervous system according ICD10, Chapter VI, codes: G00-G99**

|  |  |
| --- | --- |
| Diseases of the nervous system | G00-G99 |
| Inflammatory diseases of the central nervous system | G00-G09 |
| Systemic atrophies primarily affecting the central nervous system | G10-G13 |
| Extrapyramidal and movement disorders | G20-G26 |
| Other degenerative diseases of the nervous system | G30-G32 |
| Demyelinating diseases of the central nervous system | G35-G37 |
| Episodic and paroxysmal disorders | G40-G47 |
| Nerve, nerve root and plexus disorders | G50-G59 |
| Polyneuropathies and other disorders of the peripheral nervous system | G60-G64 |
| Diseases of myoneural junction and muscle | G70-G73 |
| Cerebral palsy and other paralytic syndromes | G80-G83 |
| Other disorders of the nervous system | G90-G99 |