HANDBOOK OF CLINICAL NEUROLOGY

VOLUME 19

TUMOURS OF THE SPINE AND SPINAL CORD

PART I

Edited by

P.J. VINKEN and G.W. BRUYN

TUMOURS OF THE SPINE AND SPINAL CORD

PART I

Edited by

P.J. VINKEN and G.W. BRUYN

Associate Editor

HAROLD L. KLAWANS

NORTH-HOLLAND PUBLISHING COMPANY · AMSTERDAM · OXFORD

AMERICAN ELSEVIER PUBLISHING CO., INC. · NEW YORK

Library of Congress Catalog Card Number: 68-8297

ISBN for the complete set: 0 7204 7200 8

ISBN North-Holland for this volume: 0 7204 7219 9

ISBN American Elsevier: 0 444 10783 5

222 illustrations, 52 tables

PUBLISHED BY:

NORTH-HOLLAND PUBLISHING COMPANY · AMSTERDAM

NORTH-HOLLAND PUBLISHING COMPANY, LTD. · OXFORD

SOLE DISTRIBUTORS FOR THE U.S.A. AND CANADA:

AMERICAN ELSEVIER PUBLISHING COMPANY, INC.

52 VANDERBILT AVENUE

NEW YORK, N.Y. 10017

PRINTED IN THE NETHERLANDS

Foreword to volumes 19 and 20

These two volumes on spinal tumours complete a series of five on space-occupying processes of the central nervous system. The initial symptoms of spinal tumours may be at least as subtle as those of cerebral tumours, even if the coordinates of functional damage, viz. those of shape, space, and time, eventually allow only a fairly stereotyped clinical phenomenology. The spinal cord, by virtue of its elongated shape, predetermines the coordinate of space to a large extent: this organ, with a length of about 45 cm and a cross-sectional area of about 2 cm², the functions of which are linked to lamellar and columnar substrata of organisation, without local functional concentrations and differentiations, is scarcely a candidate for a protean gamut of functional derangement.

It seems however, as if the spinal cord compensates for its relatively narrow range of symptoms by exploiting the potentialities of its unpaired nature. The catastrophic effect of its functional interruption, as exemplified by the spectacle of the young spinal paraplegic or tetraplegicsubject in a wheel-chair or Stryker's frame, constitutes one of the most heart-rending challenges of neurology. In the spring of 1967, one of the Editors (G.W.B.) had luncheon with the Drs Garcin, Bonduelle and Gathier at Raymond Garcin's favourite restaurant in Paris. The discussion centred around two subjects, namely the paramount, diagnostic value of history-taking and the feasibility of fundamental therapeutic progress in spinal lesions. Garcin summed up his 50 years of experience in neurology in a single sentence: 'Il n'y a qu'une chose que je reproche au bon Père et c'est de nous avoir donné qu'une seule moelle épinière'.

Other factors than its elongated shape and the fact that it is an unpaired organ confer upon the spinal cord a special brand of symptomatology. The enclosure of this organ in a bony tube which will not yield to space-occupying lesions introduces localised pressure as a major disabling element. Accordingly, the Editors decided to assign all space-occupying lesions within the vertebral canal to the category of 'spinal tumours', including even lesions that are essentially either benign or not of blastomatous nature at all. Thus, intervertebral disc prolapse stretching the spinal nerve root figures in these volumes, as well as cartilaginous exostoses and neurenteric cysts. A natural consequence of this decisions would have been also to include lesions such as epidural haematoma

or abscess. However, in order to achieve a practical demarcation of categories of lesions, the Editors have allocated spinal epidural haematoma to the volumes on injuries, well aware as they are that this haematoma is rarely, if ever, of traumatic origin. Most people would undoubtedly agree with this assignation, and would look for spinal epidural haematoma in the volumes dealing with injuries.

The final coordinate in the diagnosis, particularly in the treatment of spinal space-occupying processes, is time. It has been too often the experience of the Editors (and indubitably also of many clinical neurologists) that the main shortcoming in the practice of neurology is the lack of organisational and diagnostic alacrity in forestalling the accomplished fact of a complete cord transection, or, once it has developed, in instituting appropriate surgical treatment. This latter action is too often delayed notwithstanding the proven fact that recovery of function is impossible unless this treatment is begun within hours of the onset of the complete transectional picture. 'Anticipation is the mother of control' must be the diagnostic guideline of the neurologist in this group of decisions, and the highest level of energetic and purposeful collaboration between neurologist, neuroradiologist and neurosurgeon should be the dominating principle of therapeutic organisation.

The reader will discover in these two volumes the same basis concept of layout as followed in all the preceding volumes. The matrix of the site and the nature of the lesions underlies the pattern of each chapter. It enters into every facet of the entire field and aims to promote diagnostic acumen, since the insidious and misleading nature of spinal symptoms has proved a pitfall for many a physician. The mimicry of foramen magnum tumours and the cases of thoracic disc hernias masquerading as cases of multiple sclerosis are familiar to the student of neurology and are given a prominent place.

The first of these two volumes provides in the main general diagnostic and methodological information. Dr. Schäfer's contribution on differential diagnosis should have figured as Chapter 5, but, owing to problems in translation and printing deadlines, it has had to be moved to the end of Volume 19. The Editors would particularly like to thank Dr. Schäfer for his untiring patience and collaboration in preparing the eventual manuscript. Other contributors who deserve special mention are Dr. du Boulay of London and Dr. Pinto of Rio de Janeiro. Dr. du Boulay did not only kindly agree to be responsible for the chapter on myelography at a very late stage in the production schedule, but also submitted his manuscript within three months. Dr. Pinto deserves a particular warm word of thanks in that he completed his assignment under the somewhat unusual and trying circumstances imposed upon him by having to serve a term of confinement in prison for political reasons. He may perhaps be able to draw some comfort from the knowledge that, in our experience, imprisonment has not figured thus far in the various reasons advanced for missing an editorial deadline! We look forward to it being bettered!

In Part II special thanks are due to the Drs Odom and Wilkins who wrote two excellent chapters in fine 'Handbuch' style. Professor Nittner and Professor

Kuhlendahl likewise deserve our appreciation for having stepped, so willingly and ably, into the breach caused by contributors being unable to complete their promised assignments. We are doubly grateful to them for having been able to prepare their respective chapters, despite the demands made on them by their clinical responsibilities.

The Editors are confident that these volumes will provide the reader with comprehensive clinical information concerning a too often neglected sector of neurology. They hope to contribute, via these volumes, to the awareness of the unique vulnerability and irreplaceability of the spinal cord. If they have helped to impart to their junior colleagues the sense of urgency required in the diagnosing of spinal disease and the necessity to keep the mental image of a paraplegia or tetraplegia in mind whenever one is confronted with the possibility of a spinal lesion, the time spent and the effort expended in the preparation and execution of these two volumes will not have been in vain.

P. J. V.
G. W. B.

Acknowledgement

Several illustrations and diagrams in this volume have been obtained from other publications. Some of the original figures have been sligthly modified. In all cases reference is made to the original publications in the figure caption. The full sources can be found in the reference lists at the end of each chapter. The permission for the reproduction of this material is gratefully acknowledged.

List of Contributors

M. Joan Heffelfinger
Department of Neurosurgery and Pathology, Mayo Clinc and Mayo Foundation, Rochester, Minn. 287

J. P. W. F. Lakke
Department of Neurology, University Hospital, Groningen 91

E. C. Laterre
Neurological and Neurosurgical Clinic, Catholic University of Louvain 125

J. A. R. Lenman
Department of Medicine, University of Dundee 267

Philip Levitt
Department of Neurosurgery, New York University School of Medicine, New York, N.Y. 77

Collin S. MacCarty
Department of Neurosurgery and Pathology, Mayo Clinic and Mayo Foundation, Rochester, Minn. 287

C. Manelfe
Department of Neuroradiology, Hôpital de Purpan, Toulouse 139

Feliciano Pinto
Neurosurgical Clinic, Hospital Adventista Silvestre, Rio de Janeiro 245

Joseph Ransohoff
Department of Neurosurgery, New York University School of Medicine, New York, N.Y. 77

J. Roulleau
Department of Neuroradiology, Hôpital de Purpan, Toulouse 139

E.-R. Schäfer
Neurosurgical Clinic of the University of Göttingen 347

Hans Schliack
Neurologic Department, Steglitz Clinic, Free University of Berlin 23

Neil Spielholz
Department of Physical Medicine and Rehabilitation, New York University School of Medicine, New York, N.Y. 77

Dirk Stille
Neurologic Department, Steglitz Clinic, Free University of Berlin 23

Tapio Törmä
Neurosurgical Service, Central Hospital, University of Turku, Finland 293

H. Vogelsang
Department of Neuroradiology, Academy of Medicine, Hannover 229

Contents

CHAPTER 1

Statistical aspects of spinal cord tumors

MILTON ALTER

University of Minnesota Center for Health Sciences, Minneapolis Veterans Hospital, Minneapolis, Minn.

Clinical information about spinal cord tumors should be built upon the bedrock of precise statistics. Yet, for the most part, data about statistical aspects of spinal cord tumors are meager and biased. Information is largely derived from clinical series collected often from a single institution or from series published by different investigators using diagnostic criteria which may not be uniform. Rarely have data about spinal cord tumors been related to a well defined population so that differences between series may merely reflect differences in age, sex or ethnic distribution of the populations from which the series were drawn. These shortcomings of statistical data about spinal cord tumors must be borne in mind when considering reports of frequency, age distribution, sex preponderance, symptoms and prognosis.

FREQUENCY

One way to estimate the frequency of spinal cord tumors is to calculate the incidence in the general population. Such estimates are based on all new cases of spinal cord tumor recognized per unit time, usually a year. Estimates of incidence are presumably better than proportional statistics based on large autopsy or

TABLE 1

Spinal cord tumors.
Frequency rate per 100,000 population in selected communities.

Author	Community	Annual incidence	Prevalence	Average annual mortality
Percy et al. (1972)	Rochester, Minn.	1.3	–	–
Kurland (1958)	Rochester, Minn.	2.5	12.9	0.5
Leibowitz et al. (1971)	Israel	0.9	–	–
Gudmundsson (1970)	Iceland	1.1	10.1	0.5

References, p. 20

surgical series since not every patient with a spinal cord tumor comes to autopsy nor are all individuals in the community necessarily referred for surgery to the medical facility from which the surgical series is collected. Population studies rely upon various sources of medical information in the community and data are usually collected from all hospitals, clinics and nursing homes serving that community. Data are also gathered from medical practitioners and vital statistics are reviewed to identify patients certified as having died of a spinal tumor. Only communities with well developed medical facilities and a well defined population are suitable for incidence studies. According to the few population studies of spinal cord tumors which have been carried out (Table 1), the average annual incidence varies from 0.9 to 2.5 per 100,000 population. Thus, spinal cord tumors are uncommon and occur with considerably lower frequency than brain tumors. In Rochester, Minnesota (Kurland 1958) the age-adjusted average annual incidence rate of primary brain tumors was 7.7 compared to a rate of 2.5 per 100,000 population for spinal cord tumors. In Israel (Leibowitz et al. 1971), 90 out of 1,354 (7%) nervous system tumors involved the spinal cord. The average annual incidence was 9.8 and 0.9 per 100,000 population for primary brain and spinal cord tumors, respectively. In Iceland, Gudmundsson (1970) reported a frequency of 1.1 per 100,000 population for spinal cord tumors compared to 7.8 for primary brain tumors. In 10 selected metropolitan areas of the United States, the frequency of central nervous system tumors other than those in the brain was about 1 per 100,000 population compared to rates of under 3 to almost 6 per 100,000 for brain tumors listed by sex and race (Table 2; Dorn and Cutler 1955). Percy et al. (1972) recently extended the earlier study of central nervous system tumors in Rochester (Minn.) (Kurland 1958) and reported incidence for the 34-year period 1935–1968. Primary brain tumors occurred with an incidence of 12.5 compared to an incidence of 1.3 per 100,000 population for primary neoplasms of the spinal cord. Based on these several population studies, primary brain tumors are from 3 to 12 times more common than primary spinal cord tumors.

A second method of estimating frequency is based on the prevalence of spinal cord tumors in the community at a given time. Because of different survivorship rates for different kinds of spinal cord tumors, prevalence is a less satisfactory estimate of frequency than incidence. In Table 1, prevalence estimates of spinal cord tumors are shown for Rochester (Minn.) and Iceland. The estimates agree and suggest that about a dozen cases of spinal cord tumor per 100,000 population are living in the community at any given time. Since prevalence divided by incidence gives the average duration of a disease (P/I=D), it would appear that, on the average, patients with spinal cord tumors survive approximately 10 years. This crude estimate of duration agrees fairly well with actual observations (Slooff et al. 1964) and will be discussed in more detail later.

TABLE 2

Brain and other nervous system tumors. Age-adjusted rate per 100,000 population in 1967 by sex and race. (After Dorn and Cutler 1955.)

		Brain	Other *
Male	White	5.9	1.2
	Non-white	3.1	1.3
Female	White	3.8	1.2
	Non-white	2.7	0.9

* Including spinal cord tumors.

MORTALITY

Incidence and prevalence are measures of morbidity due to spinal cord tumors and are estimates based on living patients. Mortality represents another estimate of frequency which is based on deceased individuals. As shown in Table 1, the mortality due to spinal cord tumors was identical in Rochester and Iceland; in both it was 0.5 per 100,000 population per year. Mortality for neoplasms of the 'brain and other parts of the nervous system' was close to 5 per 100,000 population in most of

References, p. 20

the 27 countries listed in a review by Goldberg and Kurland (1962). If it is assumed that spinal cord tumors constitute about one-tenth of all nervous system tumors, then mortality due to spinal cord tumors is close to 0.5 per 100,000 population in most of these countries.

Data on mortality due to spinal cord tumors in the United States are available only from 1958 onward and are listed in Table 3. These data suggest that the mortality is uniform at least over the short period for which national statistics in the United States are available. Thus, the frequency of spinal cord tumors appears to be uniform over time and place on the basis of available information. However, Percy et al. (1972) pointed out that a high proportion of central nervous system tumors is discovered only at autopsy. In less than half of patients with known primary nervous system tumors was the diagnosis of tumor listed on the death certificate. Although this error rate for death certificates was based on both brain and spinal cord tumors, there is no reason to believe that omission of spinal cord tumors from the death certificate is less common than omission of brain tumors since many patients with spinal cord tumors die of causes unrelated to the tumor. If anything, the omission rate might be higher for spinal cord tumors than Percy et al. (1972) reported and mortality data may not be an adequate source on which to base firm ideas about frequency of cord tumors.

TYPES OF SPINAL CORD TUMOR

The lack of agreement concerning classification of tumors of the spinal cord makes it difficult to estimate the frequency of various histological types. Moreover, some series included inflammatory masses such as gummas and metastases to the spinal cord whereas other excluded these lesions. Vascular anomalies were excluded by some workers (Lombardi and Passerini 1961; Percy et al. 1972) on grounds that these were malformations and not neoplasms.

In Table 4A the frequency of various spinal cord tumors is listed by type. From these studies it appears that among spinal cord tumors, neurinomas are encountered most frequently, meningiomas are second and gliomas are third in frequency. In the glioma group especially, there is a wide variety of histological subtypes some of which may be labelled by more than one term. Ependymoma is the type of glioma which occurs with highest frequency in the spinal cord in many series (Woods and Pimenta 1944; Woltman et al. 1951; Turnbull 1962) although, among histologically verified lesions reported by Shenkin and Alpers (1944),

TABLE 3

Spinal cord tumors. Mortality and death rate per 100,000 in the United States by color and sex (1958–1967 *).

Year	White				Non-white				Total				Both	
	male		female		male		female		male		female			
	No.	rate	No.	rate	No.	rate	No.	rate	No.	rate	No.	rate	No.	rate
1958	65	0.1	56	0.1	7	0.1	5	0.1	72	0.1	61	0.1	133	0.1
1959	93	0.1	54	0.1	8	0.1	3	0.0	101	0.1	57	0.1	158	0.1
1960	59	0.1	56	0.1	5	0.1	9	0.1	64	0.1	65	0.1	129	0.1
1962	72	0.1	50	0.1	13	0.1	5	0.0	85	0.1	55	0.1	140	0.1
1963	78	0.1	45	0.1	8	0.1	10	0.1	86	0.1	55	0.1	141	0.1
1964	65	0.1	52	0.1	9	0.1	9	0.1	74	0.1	61	0.1	135	0.1
1965	62	0.1	50	0.1	12	0.1	11	0.1	74	0.1	61	0.1	135	0.1
1966	75	0.1	67	0.1	15	0.1	9	0.1	90	0.1	76	0.1	166	0.1
1967	64	0.1	44	0.0	10	0.1	6	0.0	74	0.1	50	0.0	124	0.1

* 1961 not available.

References, p. 20

astrocytoma was more common than ependymoma. Astrocytomas are far less common in the spinal cord than in the brain. The glioma group also includes oligodendrogliomas and medulloblastomas although sometimes the latter are listed separately (Mackay and Sellers

TABLE 4A

Distribution of spinal cord tumors by histologic type (%). Series including all ages.

Type	Slooff et al. (1964)	Leibowitz et al. (1971)	Rand (1963)	Oddsson (1947)
Neurilemoma	29.0	25.6	25.6	25.7
Meningioma	25.5	27.8	46.5	33.3
Glioma	22.0	12.2	11.6	20.1
Sarcoma	11.9	–	9.3	12.5
Vascular tumor	6.2	8.9	2.3	4.2
Chordoma	4.0	1.1	2.3	–
Epidermoid, dermoid, teratoma and cyst	1.4	–	2.3	3.5
Other	–	–	–	–
Unspecified	–	24.4	–	< 1
Total	100.0	100.0	100.0	100.0

TABLE 4B

Distribution of spinal cord tumors by histologic type (%). Series including only children.

Type	Ingraham and Matson (1954)		Coxe (1961)*		Hamby (1935)	
	No.	(%)	No.	(%)	No.	(%)
Neurofibroma	5	8.3	3	5.1	7	7.3
Meningioma	2	3.3	–	–	5	5.2
Glioma	21	35.0	20	33.9	23	24.0
astrocytoma	9		10			
ependymoma	1		3			
medulloblastoma	4		1			
oligodendroglioma	–		1			
neuroblastoma	6		1			
ganglioneuroma	1		3			
unspecified glioma	–		1			
Sarcoma	7	11.7	17	28.8	25	26.0
Vascular tumor	1	1.7	2	3.4	–	–
Chordoma	1	1.7	–	–	–	–
Epidermoid, dermoid, teratoma and cyst	23	38.3	16**	27.1	10	10.4
Unspecified	–	–	1	1.7	–	–
Other	–	–	–	–	26	27.1
Total	60	100.0	59	100.0	96	100.0

* 1 reticuloendotheliosis, 1 metastatic tumor and 1 syrinx excluded from this series.

** Includes 3 dermal sinuses.

References, p. 20

1968; Leibowitz et al. 1971). Other tumor types are rare. Sarcomas occur about half as often as gliomas and vascular tumors (malformations) half as often as sarcomas. Chordomas are less common than vascular tumors. In a series which includes patients of all ages, the congenital tumors account for about 2%.

Tumor types differ considerably in series based only on children. The experience of various investigators who studied children is summarized in Table 4B. The congenital tumors predominate and meningiomas are very rare.

Percy et al. (1972) showed that the frequency of various types of spinal cord tumor differs in autopsy series and in series in which the diagnosis was made while the patients were living. For example, the ratio of meningiomas to gliomas in those who came to autopsy was 3:1 whereas among those diagnosed while alive, the ratio was 0.7:1. The relatively higher frequency of meningiomas in the autopsy series is accounted for by their more benign nature; often they are not even suspected during life and are discovered incidentally post mortem.

FREQUENCY BY SITE

The frequency of tumors at various levels of the spinal cord is proportional to the length of the segments. Slooff et al. (1964) listed the following lengths: cervical 10 cm, thoracic 26 cm, lumbar 8.5 cm. The expected frequencies of lesions based on the length according to the root and vertebral level is given in Table 5 modified from Slooff et al. (1964). The proportion of spinal cord tumors which occurred at these various levels was summarized from reports in the literature by Slooff et al. (1964) and comparisons between the expected and observed frequency are given in Table 5. According to Kernohan and Sayre (1952) approximately 1% of spinal cord tumors involve multiple levels. Multiple involvement also occurs with increased frequency in neurofibromatosis. In the series of Lombardi and Passerini (1961) 10 patients out of a total of 243 with primary cord tumors had more than one tumor; six of these had multiple neurofibromatosis; one had two neurofibromas, another had three neurofibromas and two each had two meningiomas. Children tend to have more extensive involvement than adults.

Besides analyzing the frequency of tumors at various levels in the spinal cord, it is of interest to know the proportion which are intramedullary, extramedullary but intradural, and extradural. Data bearing on this point presented by

TABLE 5

Spinal cord tumors by site.

Source	Number	Cervical (%)	Thoracic (%)	Lumbosacral (%)
Denk (1932)*	713	23.5	59.5	17.0
Robineau (1932)*	64**	15.6	62.5	18.7
Bunts (1935)	36	11.4	68.6	20.0
Ingebrigtsen and Leegaard (1939)	24	25.0	50.0	25.0
Elsberg (1941)	275	23.0	56.1	20.9
Ricard et al. (1953)*	206	25.5	56.0	18.5
Lombardi and Passerini (1961)	243	17.3	64.2	18.5
Rasmussen et al. (1940)	557	18.0	54.0	28.0
Steinke (1918)	324	23.8	54.9	21.3
Expected by:				
root level		23.0	58.0	19.0
vertebral level		22.5	51.5	26.0

* Summarized from Slooff et al. (1964).
** 3.2% dorsolumbar.

References, p. 20

Steinke (1918), Rasmussen et al. (1940) and Oddsson (1947) are summarized in Table 6. Individual tumors have a proclivity for certain sites which will be discussed subsequently when selected tumors are considered in more detail.

AGE DISTRIBUTION

Surgical series (Slooff et al. 1964) suggest that the peak frequency of spinal cord tumors occurs in middle life, but Kurland (1958) challenged this notion and suggested that the frequency of nervous system tumors increases progressively with age. Kurland's conclusion has recently been supported in a more extensive study in Rochester by Percy and associates (1972). Fig. 1 (from Percy et al. 1972) shows the age distribution of brain tumors in Rochester (Minn.). Other population studies have shown a decline in frequency of brain tumors in old age.

TABLE 6

Location of spinal cord tumors, based on surgical series.

	Rasmussen et al. (1940)		Steinke (1918)		Oddsson (1947)	
	No.	(%)	No.	(%)	No.	(%)
Extradural	154	28	55	16.6	26	18.0
Intradural						
extramedullary	297	53	97	29.4	74	51.4
intramedullary	64	11	36	11.0	38	26.4
intra and extramedullary	42	8	–	–	6	4.2
Root canal and cauda	–	–	30	9.1	–	–
Vertebrae	–	–	58	17.6	–	–
Not stated	–	–	54	16.3	–	–
Total	557	100	330	100.0	144	100.0

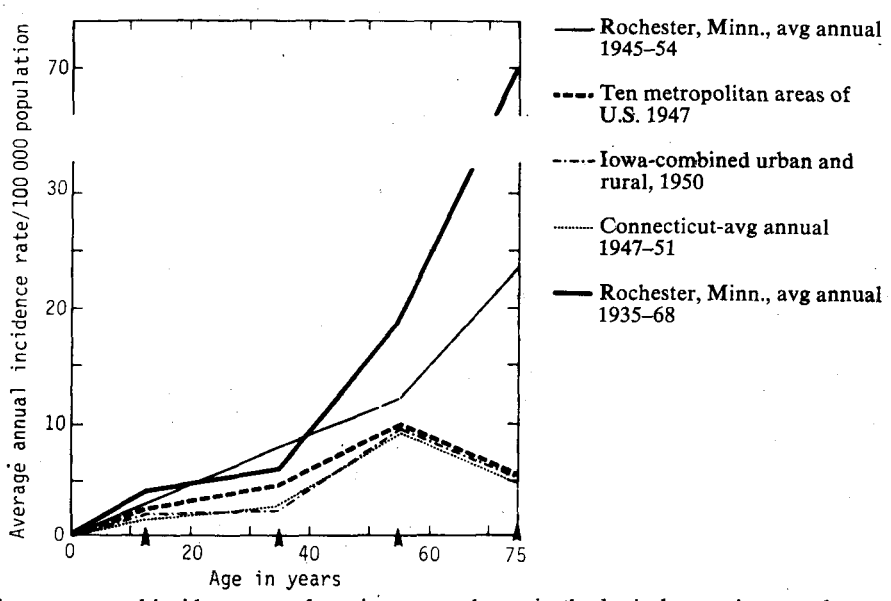

Fig. 1. Average annual incidence rates for primary neoplasms in the brain by age in several communities. (Percy et al. 1972.)

References, p. 20