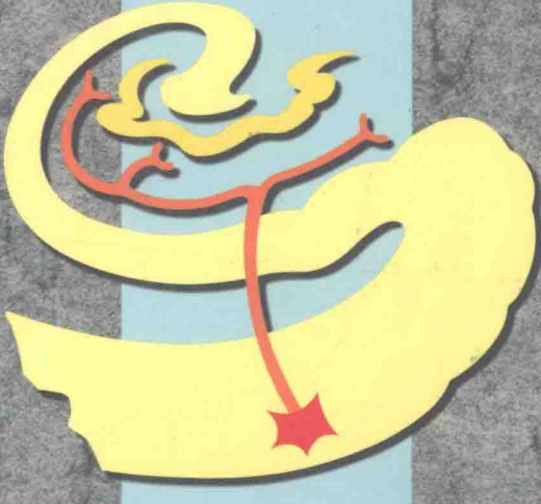
2nd Revised Edition

# Topical Diagnosis in Neurology

Anatomy · Physiology · Signs · Symptoms

Peter Duus



Translated by
R. Lindenberg

Thieme

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432 illustrations by Gerhard Spitzer

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# Preface to the Second Edition

It can be safely stated that this book has gained the reputation of being a classic. It is now referred to in many countries as "THE DUUS". This second English edition represents a revised form of the book's fourth German edition of 1987. As I went over text and illustrations again in preparing them for the second printing, I paused here and there and happily experienced the same good feelings towards this masterpiece that I had expressed in the preface to the first English edition.

This time, I am in the company of many experts who had reviewed the book. They all praise the clarity and simplicity with which the author has organized and presented the very complex and difficult subject matter of what I call *basic* neurology. They agree with the author that it should be read straight through, as one reads a novel, starting with page one. They comment on the wealth of information contained in each chapter and on the ease with which it is written. One wrote that the book is a joy to read, and another that it is difficult to become bored reading it. All praise the copious drawings by Gerhard Spitzer, Professor of Medical Graphic Arts, which are both instructive and beautiful, and recognize their share in the growing popularity of the DUUS.

Most reviewers agree with me that the book, originally written as an introduction to the broad and intricate field of *special* neurology for medical students, interns, and residents, is recommended reading also for the general practitioner and the accomplished, practicing, or researching specialist in any of the many fields dealing with the nervous system.

Some may question this, in view of the incredible, almost explosive advances made in the various modes of diagnostic imaging during the last few years. One may ask whether it still pays to learn all the minutiae of anatomy and physiology and the signs and symptoms of every segment of the nervous system. I believe it does! I believe it is simply elementary for any educated dealing with the nervous system to know at least as much about it as what "THE DUUS" offers. This holds true, without question, for the daily practice of neurology and psychiatry, and equally applies to that of neuroimaging and neuropathology, and of the most recent field of endeavor, psychoneuroimmunology.

*June 1989*
*Cockeysville, Maryland*

*Richard Lindenberg*

# Foreword

There is the danger that diagnosis through clinical observations is dying out in neurology and is being replaced by diagnosis through laboratory tests. This is the course of clinical neurology in Germany, at least. Although every text on neurology deals with topical diagnosis, it is usually given not much attention. Diagnosis by laboratory methods, even if as sophisticated as serial angiograms, scintigrams and emi-scanners, is yielding little or no information in many disease states in comparison with the diversity offered by the topical diagnostic analysis of brain and spinal cord.

The topical diagnostics of the central nervous system, as it was taught by Robert Bing, did not remain at its 1930 level. It requires familiarity with much new knowledge in neuroanatomy and neurophysiology, and with transmitter neurochemistry. The neurone theory, confirmed by electron microscopy of brain and spinal cord, constitutes the theoretical foundation of modern topical diagnosis. An injury of a neuron, regardless of whether it involves the perikarion or the axon down to its synaptic contacts, will always produce the same sign, according to this theory. The neurone theory represents the principle of order in the topical diagnosis of the central nervous system.

The present "Topical Diagnosis in Neurology" by Professor Peter Duus, an experienced neurologist and pupil of Karl Kleist, satisfies a long existing need on the book market. It provides the cogently required diagnostic implement for the practicing neurologist as well as for general practicioners and specialists in other fields, who happen to be interested in neurologic problems. It provides a balance to technical and laboratory methods for the clinical neurologist and is particularly interesting because diverse function systems producing different signs of deficits are packed very closely together in the central nervous system.

Neurologic diagnosis without topical diagnosis will always be inadequate and is frequently incorrect. The coordination of certain functional deficits to certain neuronal systems constitutes a fountain of knowledge in brain research, which cannot be overvalued and cannot be replaced by animal experiments.

For the welfare of the patients, I wish that this book of Peter Duus will enjoy the widest circulation and will be consulted over and over again.

*Frankfurt/Main* *Rolf Hassler*

# Preface to the First Edition

"If clinical neurological work in the future is to bring results of value, it is essential that the neurologist understands the major principles in the organization of the nervous system, and that he has a fair knowledge of its structure and function." A. Brodal

It is the purpose of this small book on topical neurologic diagnostics to acquaint students, interns and residents with the specialty of neurology by keeping the text concise and by using the greatest possible number of illustrations for conveying much information. Perhaps, the book offers valuable suggestions also to the practicing physician interested in neurology.

A well-based knowledge of the structural and functional relationships within the nervous system is requisite for the understanding of signs, symptoms, and syndromes of the various diseases and injuries of the nervous system and for bringing them into proper perspective for diagnosis.

Differential diagnosis is based on such knowledge and also on data collected from pointed anamnestic questioning and on the results of physical and neurologic examination conducted in search for focal as well as neighborhood signs. The conclusions drawn after differential diagnostic elaborations determine which further procedures should be used and which of the various technical diagnostic tools can most effectively be applied. The result of one or the other technical examination may corroborate what was tentatively diagnosed before or may suggest to employ additional methods.

The use of technical diagnostic procedures alone is liable to fail without carefully collecting anamnestic data and performing a routine neurologic checkup. This is particularly true when dealing with an incipient disease process. One reason for neurology being so fascinating and attractive is the possibility to deliberate on differential diagnosis just by analysing anamnestic data and basic clinical findings.

Presenting an overview of the very large field of neurology within the framework of a small book required not always easy decisions. In order to keep the descriptive text concise, illustrations had to be more numerous than usual and had to be as instructive as possible. Since the material to be presented had to be selective, certain subjects, however important, could only be touched upon or had to be omitted. These concessions notwithstanding, it is hoped that the description of those structural and functional features of the nervous system, which are important to know in the daily practice of neurology, have come out clearly and comprehensibly.

To illustrate the book so richly required tireless assistance of an expert in graphic arts, very astute in medical matters. He is Mr. Gerhard Spitzer of Frankfurt/Main. I am grateful to him for his most pleasant cooperation, his support and, particularly, for the patience he had with me.

I also want to thank very much Professor Dr. Rolf Hassler, Max Planck Institute for Brain Research, Frankfurt/Main, for reviewing text and illustrations in spite of being burdened by his own work. He gave me important suggestions and valuable stimuli.

*Frankfurt/Main, July 1976* *Peter Duus*

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# 1 Sensory System

## Receptors

Receptors are specialized sensory organs capable of registering certain changes in their vicinity and within the organism and of transmitting these stimuli as impulses. They are the end-organs of afferent nerve fibers. One can subdivide them according to their functions into *exteroceptors,* which tell the body what is going on in its ambient environments and *teleceptors,* such as those in eyes and ears, which register stimuli originating in the more distant environment.

*Proprioceptors,* among them the receptors in the labyrinth, provide information about the position and movement of the head in space, about tension in muscles and tendons, about the position of joints, about muscular strength, and about other movements and positions of the body. Finally, there are *entero-* and *visceroceptors*, which report on events taking place within the organism. They are *osmo-*, *chemo-*, *baroceptors*, and others. For the various receptors to react, stimuli must be appropriate.

*Skin receptors* will be discussed first (Fig. 1.**1**). They are subdivided into *mechanoceptors* (touch, pressure), *thermoceptors* (cold, heat), and *nociceptors* (pain). These receptors are abundant in the skin, particularly between the epidermis and connective tissue. Consequently, the skin may be looked upon as a sensory organ covering the entire surface of the body.

Skin receptors consist of two large groups: (1) free nerve endings and (2) encapsulated end-organs. *Free nerve endings* occur in the spaces between the epidermal cells and between structures of neural origin, such as Merkel's *tactile menisci* (menisci tactus). Free nerve endings are present over almost the entire surface of the body and transmit pain and temperature impulses produced by injury to the cells. The tactile menisci are located mainly at the fingertips and react upon active touching or passively being touched.

*Hair cuffs* take an intermediary position. They are present wherever there is hairy skin, and transmit touch stimuli. Meissner's *touch corpuscles* (corpuscula tactus) are present only in hairless skin, such as the palms of the hands and soles of the feet (as well as the lips, tip of the tongue, and genital mucosa). They are very sensitive to active and passive touch. The *Vater-Pacini lamellar corpuscles* (corpuscula lamellosa) are situated in deeper layers of the skin, particularly between the cutis and subcutis. They transmit pressure sensations. *Krause's corpuscles* (corpuscula bulboidea) have been considered cold receptors, and *Ruffini's corpuscles* (corpuscula lamellosa), receptors of warmth.

This is now questioned. The free nerve endings are also capable of registering temperature. The cornea, for example, has only free nerve endings, and they pick up cold as well as heat.

Aside from the receptors mentioned, there is a variety of other receptors in the skin, the functions of which have not been clarified.

A second group of receptors consists of those that are located in deeper tissues of the body: in muscles, tendons, fasciae, and joints (Fig. 1.**2**).

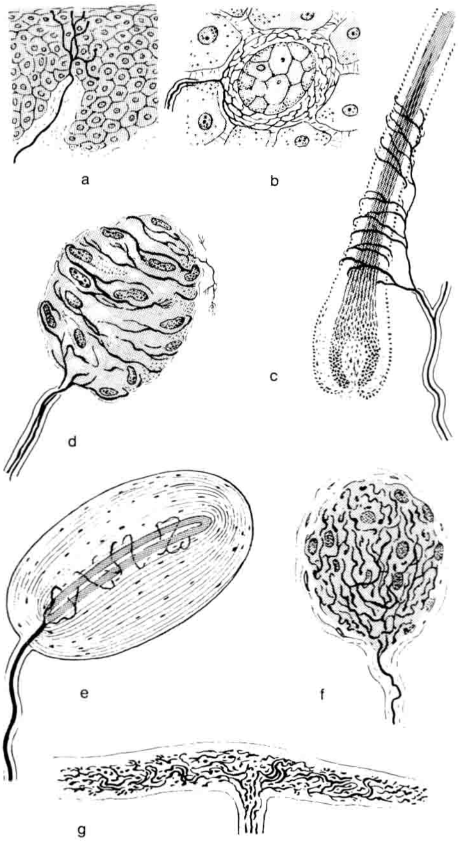


Fig. 1.**1** Endings of afferent nerve fibers (receptors) in the skin. **a** Free ending (pain, temperature); **b** Merkel's tactile meniscus; **c** hair cuff (touch); **d** Meissner's touch corpuscle; **e** Vater-Pacini's corpuscle (lamellated corpuscle; (pressure); **f** Krause's bulboid corpuscle (cold?); **g** Ruffini's corpuscle (heat?).


Fig. 1.**2** Receptors in muscles, tendons and fasciae. **a** Anulospiral ending of muscle spindle (stretching); **b** Golgi tendon organ (tension); **c** Golgi-Mazzoni's corpuscle (pressure).

*Muscle receptors* consist of several types, the most important of which are the *neuromuscular spindles*. They respond to passive stretching of the muscle and are responsible for the *stretch* or *myotactic reflex*. These thin, spindle-shaped structures are ensheathed by a layer of connective tissue and are located between striated muscle fibers of the skeletal musculature. They contain 3 to 10 very thin striated fibers called *intrafusal muscle fibers* (*fusus* means spindle) in contrast to the other or *extrafusal fibers*. The polar endings of the connective tissue capsules are tied to the diffuse connective tissue stroma that invests fibers, fascicles, and the entire muscle. In this way the spindles participate in the movements of the muscle. Afferent fibers, called *anulospiral endings* or *primary endings*, are spun around the middle of a muscle spindle. These fibers have rather thick myelin sheaths and belong to the fastest-conducting fiber group the so-called Ia fibers. The equatorial, noncontractile portion of a spindle contains 40 to 50 small nuclei in so-called *nuclear-bag* *fibers*. Attached to them are *nuclear-chain fibers* each harboring a row of individual nuclei. For more detail, see pages 7–14 (monosynaptic proprioceptive reflex and polysynaptic reflexes).

The *tendon organs of Golgi* are delicate nerve endings or branches of thickly myelinated nerve fibers that are wrapped around groups of collagenous tendon fibers. They are surrounded by a connective tissue capsule, are located at the transitional area between tendon and muscle, and are arranged in series with the muscle fibers. Like muscle spindles, they respond to tensile stimuli, but their threshold is higher (see Fig. 1.**10**).

In addition to muscle spindles and tendon organs of Golgi, there are still other types of receptors in this region that transmit pressure, pain, and other stimuli. The *Vater-Pacini lamellar corpuscles*, *corpuscles of Golgi-Mazzoni*, and *terminal nerve endings* are some of them.

All these receptors in skin and deeper tissues are attached to a collateral of an axon. Several axonal collaterals converge toward the axon of a sensory neuron. Every cutaneous stimulus impinging on the skin activates not merely one but several types of receptors. The sum total of stimuli is transmitted to the central organ as an impulse of varying velocity.

The encapsulated, more differentiated terminal corpuscles likely transmit epicritic qualities, such as light touch, discrimination, vibration, and pressure. The free nerve endings are probably responsible for transmitting protopathic qualities, such as differences in pain or temperature.

Receptors are the peripheral endings of afferent nerve fibers, which are the peripheral processes of *pseudounipolar spinal ganglion neurons*. Each neuron of the ganglion gives off a short axon, which soon branches like a T. One branch runs to the periphery, joining the receptors. The other branch connects, via the posterior root, with the spinal cord, in which it proceeds in different directions, depending on the quality of the sensory impulse it carries (see Fig. 1.**19**).

## The Peripheral Nerve

A nerve consists of one or more bundles of nerve fibers (axons). A nerve of medium size may contain thousands upon thousands of nerve fibers, some unmyelinated and others surrounded by myelin sheaths of different thickness. Fig. 1.**3** shows a nerve on cross

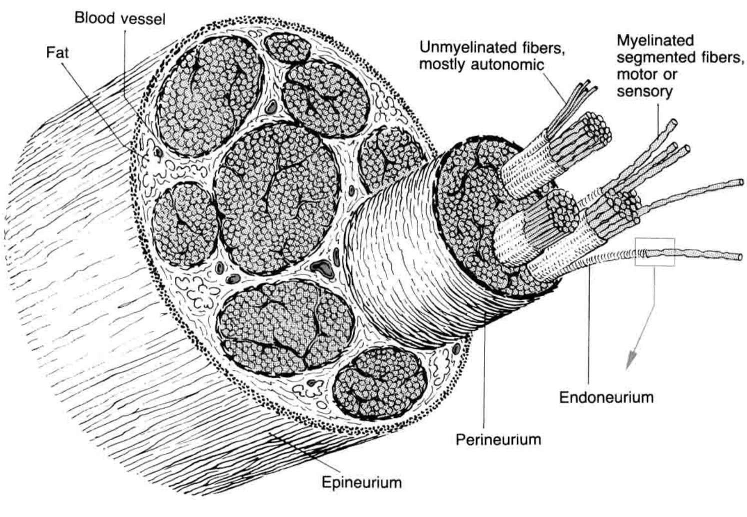


Fig. 1.**3** Cross-section of a peripheral mixed nerve.

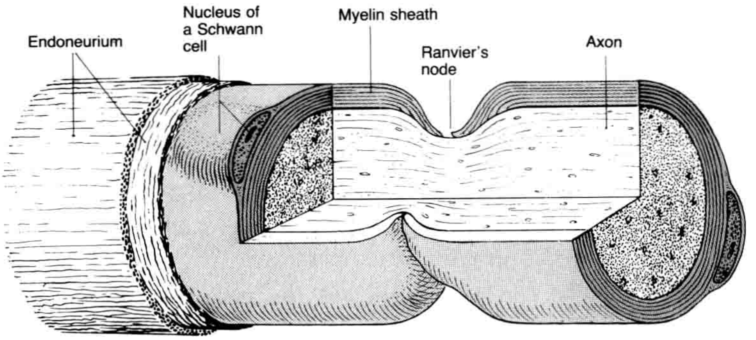


Fig. 1.4 Myelinated nerve fiber. Enlargement of fiber passing through rectangle in Figure 1.3.

section. Fig. 1.4 illustrates a single myelinated nerve fiber, cut crosswise and lengthwise, and shows that the centrally located axon is surrounded by a sheath of myelin (myelin is a mixture of lipids and protein). The nuclei of two Schwann cells can be seen. According to electron micrographs, the surface membranes of these cells are spirally wrapped around the axon, forming multiple layers enclosing lamellae of myelin, which are part of the Schwann cells. The myelin sheaths may be thought of as layers of insulating material.

The sheaths of Schwann and the myelin they contain are interrupted at 1 to 2 mm intervals by ring-shaped constrictions called *nodes of Ranvier*. These nodes play an important role in the propagation of stimulus effects from receptor to spinal cord or vice versa by facilitating fast conduction of the impulses through saltatory conduction of action potentials. The thicker the myelin sheaths, the faster the nerve fiber conducts. Both myelinated and unmyelinated or poorly myelinated fibers are surrounded by the protoplasmic membranes of the Schwann cells, only one of these cells serving the nerve fiber segment between two nodes of Ranvier.

The Schwann cells are enveloped by a layer of connective tissue, the *endoneurium*. The connective tissue surrounding several bundles of nerve fibers is called the *perineurium* and the one wrapped around larger nerves, the *epineurium*. These connective tissue coverings protect the nerve from mechanical injury and direct contact with nerve-damaging agents. The connective tissue carries the blood vessels nourishing the nerve fibers.

The peripheral nerve contains afferent as well as efferent, myelinated as well as unmyelinated, and somatic as well as vegetative or autonomic fibers. The somatic fibers connect receptors with the spinal cord, and the motor cells of the anterior horns with the musculature. The autonomic fibers are also both afferent and efferent and innervate the viscera, blood vessels, and glands.

Somatic and autonomic fibers, whether afferent or efferent, do not run in separate bundles within a mixed nerve. They are intermingled until they approach the point of destination. Then they separate again, as nerves for skin, muscles, joints, and viscera.

The nerve fibers are classified according to the thickness of their myelin sheaths and the velocity of their conduction. Table 1.1 provides examples.

The *posterior roots* contain only afferent nerve fibers. All impulses originating in receptors in skin, muscles, joints, and internal organs have to pass through the posterior roots to enter the spinal cord. *These afferent fibers are the central branches of the pseudounipolar spinal ganglion cells.* The *impulses are not switched or transferred to neurons of the spinal ganglia.*

Table 1.**1** Classification of Nerve Fibers According to Thickness of Myelin Sheaths and Velocity of Conduction.

| Type of Fiber | Diameter (μ) | Velocity (m/sec) |
|---|---|---|
| Ia fibers (A, α)<br>From anulospiral endings | Approx. 17 | 70–120 |
| Ib fibers (A, α)<br>From tendon organs of Golgi | Approx. 16 | 70–100 |
| II fibers (A, β, and γ)<br>From flower-spray endings and Merkel's touch menisci | Approx. 8 | 15–40 |
| III fibers (A, δ)<br>Pain, temperature, pressure | Approx. 3 | 5–15 |
| IV or C fibers<br>Pain, temperature, heavy touch | Approx. 0.2–1 | 0.2–2 |

The nerve fibers transmitting impulses from the various sensory receptors are intermingled in the peripheral nerve. As the nerve approaches the spinal ganglion, the fibers separate into groups according to their specific functions and take definite positions within the dorsal root (Fig. 1.**5**). The nerve fibers that originate in the neuromuscular spindles

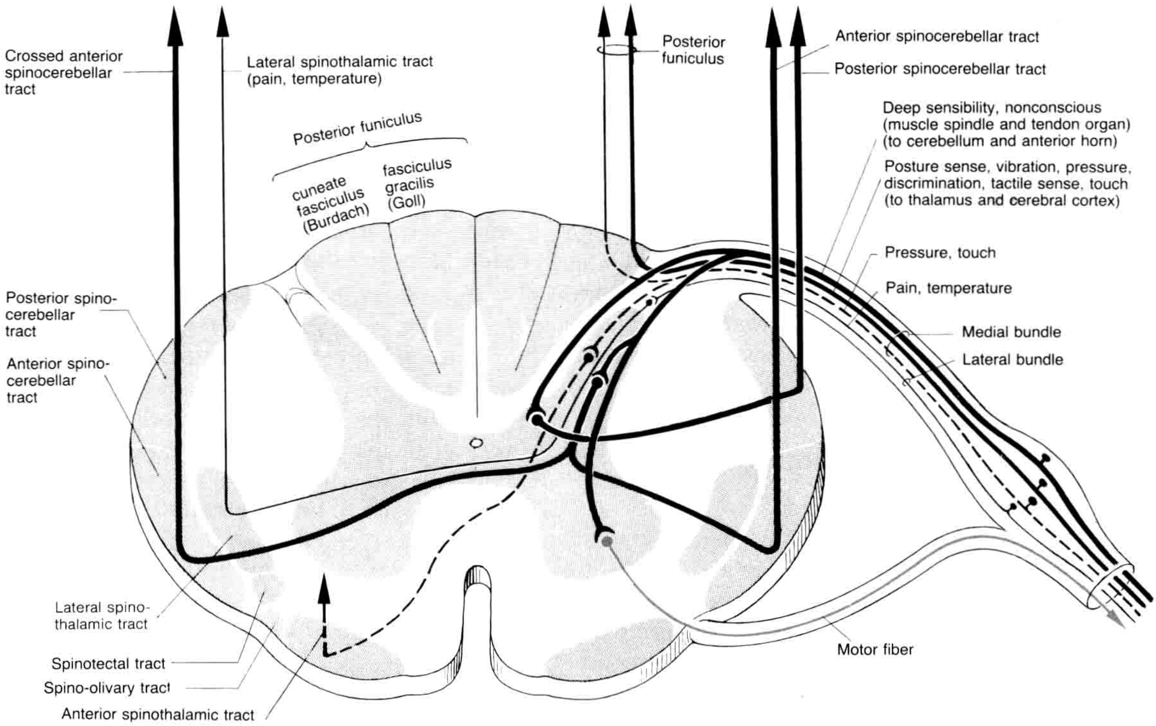


Fig. 1.**5** Course of posterior root fibers in spinal cord.

and have the thickest myelin sheaths occupy the most medial part of the root. The mid-portion of the root is taken up by fibers that derive from encapsulated receptors and transmit, among other sensations, those of touch, vibration, pressure, and discrimination. The most lateral fibers are the ones that are almost unmyelinated and carry pain and temperature impulses.

The nerve fibers with the thickest myelin sheaths conduct deep sensibility (*proprioception*). Only some of the impulses coming from muscles, joints, fasciae, and other tissues reach the level of awareness; most serve the automatic control of motor activity needed for walking and standing.

Upon passing through the *entrance zone* of the posterior roots into the spinal cord, the individual fibers divide into numerous collaterals, which secure synaptic connections with other neurons in the spinal cord. Fig. 1.**5** shows that the nerve fibers join different tracts within the cord, depending on the sensory modality they serve. It should be noted that all afferent fibers when passing through the entrance zone of the posterior root, also called the Redlich-Obersteiner area, become momentarily devoid of myelin sheaths. The transition from peripheral to central nerve fiber thus is rather abrupt. Schwann cells characteristic of the peripheral nerve cease to exist, and oligodendrocytes take over. This physiological lack of myelin in the transitional area is said to render the nerve fibers vulnerable to disease, such as tabes dorsalis.

## Neurons of the Central Nervous System

We must briefly discuss the neurons of the central nervous system before we describe the further course of the fibers that carry impulses from diverse sensory modalities into the spinal cord via spinal ganglia and posterior roots.

Fig. 1.**6** shows the afferent fiber of a pseudounipolar neuron of the spinal ganglion forming a simple *monosynaptic reflex* arc with a highly specialized motor neuron in the anterior horn of the spinal cord.

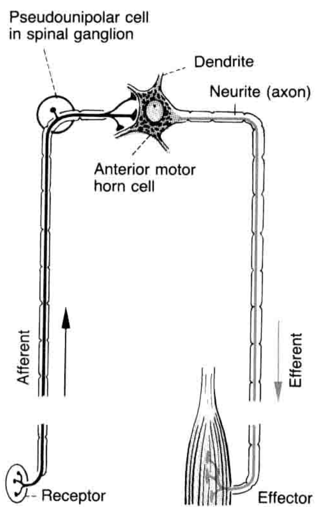


Fig. 1.**6** Simple monosynaptic reflex arc.

The structure of these neurons is so complicated that we can give only a brief description. The body or *perikaryon* of a cell of this type has numerous processes of different lengths. One of them is particularly long and carries the discharges of the cell to the periphery. It is called the *axon* or *neurite*. The others are shorter, branch extensively, and are called *dendrites*.

The neurons produce and conduct action potentials. A neuron can transmit excitation to another neuron via one or multiple points of contact, or *synapses*. Synapses are separated from the surface of the other cell by a very small space. As an excitation reaches the synapse (presynaptic), a transmitter substance is set free into the space and either enhances (acetylcholine) or inhibits ($\gamma$-aminobutyric acid) the postsynaptic element of the other neuron.

A single nerve cell receives impulses not from only one or two neurons, but from many and even thousands of neurons. A great number of *boutons terminaux* (synaptic end-feet) are attached to the outside of the