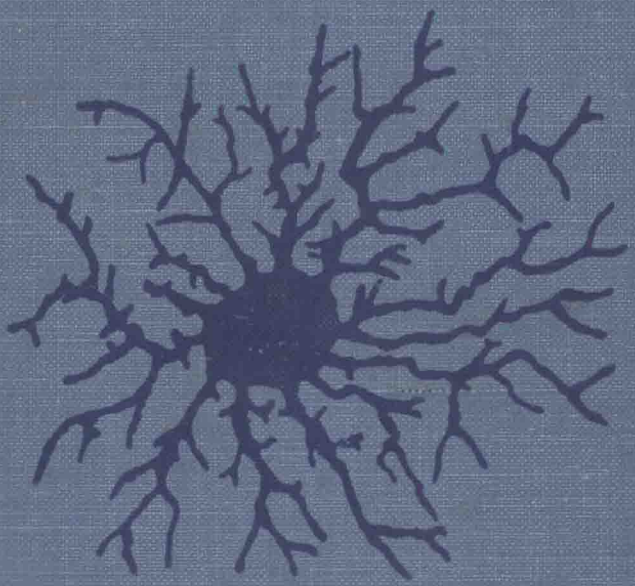
PEDIATRIC NEUROLOGIC NURSING

BARBARA LANG CONWAY



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with 102 *illustrations*

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PEDIATRIC NEUROLOGIC NURSING

TO
MY FAMILY

Preface

This book is the culmination of 10 years' experience in the nursing field; as a clinical specialist I was constantly asking questions that were frequently not entirely answered by one reference or individual in the context of my needs as a nurse. Hopefully, this book merges data from several disciplines to provide the experienced nurse with information underlying the full range of practice.

The book begins with embryology because the gestational period supplies many answers to problems that arise later. Functional physiology is an overview of neurologic operations. Chapter 3 is an overview of development and psychosocial implications, since these factors have a great bearing on the functional capacities. Each of the other chapters is designed to include the various "ball parks" that the nurse and neurologist/pediatrician consider as they perform a gross assessment of a neurologic problem.

Once the general category is known, specific definition of the disorder is appropriate. References are designed to provide further reading on subjects in the book. Actual descriptions of nursing management are limited, since their addition would make the book too lengthy and details on these procedures may be gleaned from Carini and Owens: *Neurologic and Neurosurgical Nursing* or other references given at the end of each chapter. The book is designed to supplement material learned as a part of basic neurologic nursing care.

This book is without a doubt the largest project I have ever tackled. It has taken two years to write. The book would have been impossible without the assistance of many friends and the support, love, and endurance that my family extended.

To Dr. Jane Kordana, Dr. Melvin Greer, and Dr. John Ross, who spent many hours tutoring me, I wish to express my gratitude. Many other faculty members at the University of Florida also had a great impact on my learning experiences, especially Dr. Pauline Barton, Dr. W. Whitner, Carol Taylor, Dr. Carol Bradshaw, Dr. W. W. Purkey, and Dorothy M. Smith. Some of these individuals are still at the University, while others have moved on to other institutions.

During the preparation of the manuscript I was employed at Variety Children's Hospital, where the entire nursing office gave me great encouragement in the project and suggested a number of references that might be helpful. My special thanks to Donna Thaler, Pat Eaton, Kay McLean, Carol Drozdowicz, Dorothy Thomas, Sue Cox, Erica Causey, Barbara Kriston, Kay Levine, Ruth McDowell, Betty Ducsay, Elsie Damien, Francisca Reed, Barbara Aissen, Judy Skinner, Clara Meadows, Doris Sitton, and Edith Witte. I would also like to thank Jean Vandergrift and Charlotte Dison for allowing me to utilize their research materials.

Dr. Robert Cullen, Dr. Danilio Duenas, Dr. R. Bejar, Dr. Pedro Lopez, Dr. Nora Oldham, Dr. Sergio Delamerens, Dr. Carol Hersh, and others were instrumental in answering the many questions that arose as the manuscript developed. Thanks also go to Hilda M. Lang for reading the manuscript. Marita Bitans, the illustrator, should be commended, particularly since all directions and consultation were limited to phone conversations. Billie Erlich, for her expertise and friendship, and Joyce Dorner, my clinical specialist consultant and best friend, deserve my very personal gratitude. The efforts of many others must remain unmentioned because of space limitations.

Barbara Lang Conway

Contents

1 *Embryonic development*

As a result of the knowledge explosion that has occurred in recent years, professionals are learning more about the mysteries of the conception, gestation, and birth of human beings than ever before. While many aspects of the process of embryogenesis are yet unknown, a thorough study of prenatal growth and development has proved invaluable as a basis for predicting, preventing, and treating many pathophysiologic conditions.

In recent years the morbidity and mortality rates for pregnant women have decreased so markedly that the major focus of medicine has shifted to the fetus. Definitive assessments and histories assist the practitioner in detecting inadequate intrauterine environments that may result in prematurity, retarded growth, or untoward outcomes during or subsequent to the birth process.[1,2]

FUNDAMENTALS

In reviewing the preliminary steps of embryonic growth, one should consider the complexities that surround the process of conception. Conception is dependent on the proper timing and acceptable physiologic functioning of mature male and female reproductive systems. Successful conception is also related to a combination of various factors, including social, cultural, and psychologic events. The exact interaction of these factors is not known and varies with different individuals and subcultures.

The stage is set when the ovum is released from the ovary into the fallopian tube and begins a 10-day journey to the uterus. Conception occurs when the ovum is approximately one third of the distance through the fallopian tube. As one of the millions of sperms available from a single ejaculation penetrates the ovum, a zygote is formed.

As the zygote continues to mature, it is subjected to a process known as morphogenesis, which consists of three fundamental phases: growth, cell differentiation, and development of relative movement. This chapter deals principally with the growth phase.

Growth

Growth, an everyday word, takes on a specific meaning when related to ontogeny. In this instance it is defined as increasing size and mass as a result of protoplasmic and extracellular material synthesis, a process essential to the production of specific tissue components. Therefore growth that occurs as a direct outcome of accommodating consumed foodstuffs or fluids is not a part of this particular definition.

Tissue growth may be classified as multiplicative (increase in cell number), auxetic (increase in cell size), accretionary (increase in intercellular material), or a combination of these three.

Multiplicative process

Multiplicative growth begins when the zygote is formed through mitotic cell division. This repetitive process varies in different types of tissues. In some tissues multiplicative growth is cyclic throughout life. Examples of cells that are constantly dying and being replaced are contained in the epidermis and the intestinal epithelium. In contrast, oocytes and neurons are formed during embryonic development and are never replaced or regenerated.

A great deal of research has been done and is currently taking place in the study of the vast differences to be found in the various types of

tissue growth. Much emphasis is being placed on ascertaining the precise mechanisms and interactions that regulate the production and or cessation of multiplicative growth.

General influences on this process incorporate factors such as genetic influence, nutritional input, primary organizer region, and endocrine, thermal, photic, and purely mechanical determinants. Specific cell growth, in relation to a particular tissue's needs and functions, is probably regulated by chalones, inhibitory secretions produced by the tissue itself to control mitotic cell division.

Auxetic process

Auxetic growth is a deoxyribonucleic acid (DNA)–controlled increase in cellular size. DNA is present in the diploid nuclei of each cell in a given quantity. This quantity determines production capacities for replacing the protein that is constantly being broken down during the cellular processes taking place throughout the life of the cell.

Auxetic growth ceases at the limit of the cell's optimum potential to grow and develop. However, the nucleus of some cell types that have attained this maximum auxetic growth potential may divide to allow for further growth. In other instances, such as in neuronal growth, the single diploid nucleus (of the neuron) is aided by the surrounding glial cells, which provide additional metabolic and nuclear material for further neuronal growth. This mechanism allows some neurons to expand to an extremely large size and volume postnatally. Conversely, in other cells, such as the granule cells of the cerebellar cortex, ongoing multiplicative growth results in a diminished cell volume.

Accretionary process

Accretionary growth is the process whereby an increase occurs in the quantity of structural intercellular material in a given tissue. Examples of tissues undergoing this type of growth include fibrous connective tissue, bone, tendon, cartilage, joint capsules, and the cornea.

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In reviewing various aspects of embryonic growth, one should recognize that all of these growth processes—multiplicative, auxetic, and accretionary—are combined in unique ways consistent with cellular differentiation, form and size, function, age, and regenerative processes characteristic of each particular body part. An integral part of growth, even in embryonic development, is cell death and removal. This phase is imperative not only to local tissue balance, but also to the degenerative process required in the progression from rudimentary stages of growth to maturity.[3]

Cell differentiation

Regulatory forces that influence the growth of various cells, which combine to form functioning tissues, remain the key focus of reseachers today. In the past scientists have argued over two primary positions of understanding cellular differentiation: preformation and epigenesis. Those that favored the preformation concept believed that all essential body parts were present in minute form in the zygote and that growth merely increased their size and final development so that they could be visualized. Epigeneticists believed that organ development represented new formations. Both schools of thought have more or less been incorporated into the current viewpoint, which is that cytoplasmic informational content and nuclear genetic mechanisms are the "preformants" that control the growth and development, or "epigenesis," of body structures.[4]

The key to understanding cellular differentiation is twofold: (1) Basic to all cellular components is protein. Protein is also the catalytic agent that constitutes the myriad of enzymes inextricably involved in controlling the sequence of energetic and structural changes that are fundamental to organismic development and viability. (2) The control complexes that regulate protein synthesis include an elaborate system of chemical "messengers" that involve relays both intercellularly and intracellularly. However, the master control system that determines tissue growth and development may be explained through the principles of primary organization and genetic control.

Primary organization

The concept of primary organization has evolved from numerous experiments, wherein

tissues have been transplanted and monitored for subsequent effects on growth. Results show that some tissue in the blastula, a cellular mass that floats freely in the uterus by the fifth gestational day, has an early predisposition for self-differentiation and a set course, whereas the surrounding tissue has a dependent differentiation. Regions of the blastula that exhibit dependent differentiation are pluripotent, that is, they possess the ability to become several different tissues in response to the growth of the adjacent inductive, or self-differentiating, tissue, at least in the earliest growth stages.

The most significant tissue in the early stages of self-differentiation is that of the dorsal blastopore region, since its powerful inductive abilities result in the formation of the embryonic axis and its neighboring tissues. Because of these qualities the dorsal blastopore region is called the primary organizer region. Later other subordinate organizing regions are evident in the functional structure of sequential chordate development.

Communication between inductive and dependent areas is effected through intracellular and intercellular chemical messengers, whose exact constitution has not yet been identified.

Genetic influences

The second major influence is that exerted by genetic controls. Even though the mechanisms of operation remain in dispute, a brief review of the theoretical processes is warranted.

Genetic codes are contained in the chromosomes, a double strand of DNA found in the cell nucleus. The amino acid configurations that make up DNA differ slightly in various genes. However, all genes are attached within a single chromosome. Basically, genes control enzyme synthesis, which determines body metabolism by the regulation of ribonucleic acid (RNA).

Hypothetically, DNA loci are found in functional groups called operons. Operons contain structural genes, a regulator gene, and an operant segment. Structural genes are programmed to specify polypeptides, simple amino acid molecules that form either structural or enzymatic proteins. However, the production or inactivation of these structural genes relies on whether their operant segment is free to function or is inhibited by a repressor protein that its specific regulator gene has produced (Fig. 1-1).

Repressor protein production by the regulator gene is directed by chemical substances from the *blastomeres* and the *embryonic inducers,* as

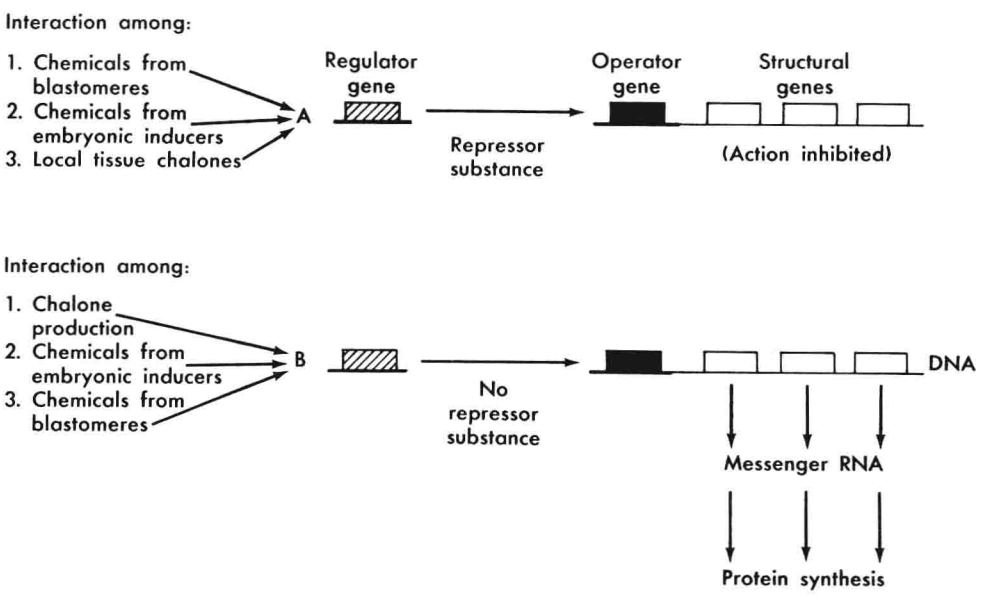

Fig. 1-1. Interactions among regulator, operant, and structural genes.

mentioned in the discussion of regional organizers. These local tissue substances that enhance repression of activity include the previously mentioned *chalones,* the balancing force between circulating hormones and the local tissue cell genome.

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In summary, sequential growth and cell differentiation are dependent on a balanced, specified pattern for the operation and inhibition of operons and effective chemical interactions of *inducers, circulating hormones, repressors, corepressors,* and *local tissue chalones.* Extrachromosomal nucleic acids possibly influence this process, but further research is needed to confirm their role.[5]

Development of relative movement

Relative movement involves selective interactions among tangential cellular surfaces. Through this surface relationship and subsequent changes in peripheral form, the formation of cell masses, groupings, and invaginations integral to embryonic development is possible. Another related concept is that of contact guidance, which is best demonstrated by the regeneration of nerves on Schwann cell surfaces (neurilemma cells) and the growth of new nerve fibers along the paths of preceding nerve fibers.

HEREDITARY PROCESSES

Understanding the inherited abnormalities that comprise large groups of childhood disorders is based on the principles of cell division and inheritance presented here.

Spermatogenesis

Spermatogenesis occurs in the seminiferous tubules of the testes after sexual maturity is attained. Within 75 days the spermatogonia complete the first and second meiotic divisions and emerge as four mature sperm cells, which possess the full complement of 23 chromosomes and the ability to move spontaneously (Fig. 1-2, *A*).

Meiosis

Of the 6 million primary oocytes originally produced in the ovary, only approximately 400 will have the potential to be fertilized during the reproductive years.

First these cells undergo the prophase of the

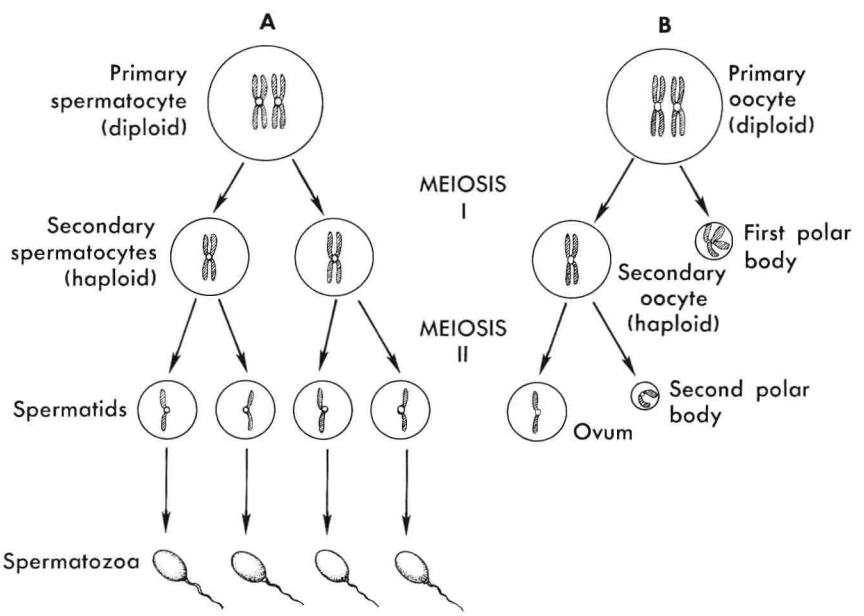

Fig. 1-2. Meiotic division of sperm, **A,** and ovum, **B.**

first meiotic division prenatally. Then each oocyte remains dormant until ovulation, when it matures along with the ovarian follicle. Prior to ovulation the first meiotic division is continued to completion.

Ovulation marks the beginning of the second meiotic division, which terminates at the time of fertilization. In the final division, the telophase, the predominant amount of cytoplasm is contained in one mature ovum, while other daughter cells remain small and unable to reproduce (Fig. 1-2, *B*).[6]

Fertilization

Once the spermatozoon penetrates the ovum, the head and neck remain within the ovum, while the midsection and the tail are detached and discarded. The head then swells into a pronucleus that gravitates toward the center of the ovum to combine with the female pronucleus. Both nuclei duplicate their respective DNA material. Two centrioles appear and chromosomes of both pronuclei align themselves on the spindle between the centrioles, where they each contribute 23 corresponding chromosomes (Fig. 1-3). The nuclei divide longitudinally, and each set migrates to the opposite pole of the cell. A deepening external groove appears on the cell as the zygote enters the developmental phase known as cleavage or segmentation.

Genes and chromosomes

Gregor Mendel initiated the science of classical genetics with a paper prepared in 1866 on factors, which were later identified as genes. These genes are in the chromosomes of all somatic cells. Each corresponding gene, or allele, has a particular location in its chromosome. Genes are ultimately transferred to future generations via the chromosomes of the ovum and spermatozoon.

At the time of fertilization, each parent normally contributes 22 autosomes and 1 sex chromosome. There are exceptions that result in

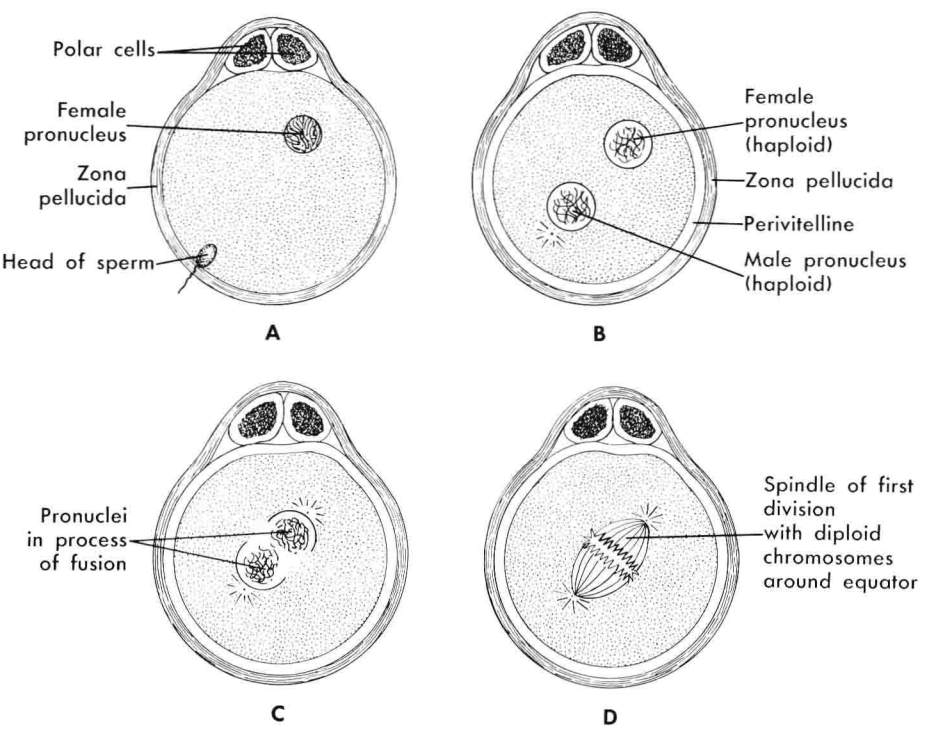

Fig. 1-3. Fertilization of ovum. **A,** Sperm penetrates ovum. **B,** Pronuclei gravitate toward center of ovum. **C,** Pronuclei fuse. **D,** Appearance of two centrioles and alignment of chromosomes around equator.

cases of genetic and metabolic disorders, which will be discussed in Chapter 6. Females contribute only the X chromosome, whereas males may offer either an X or a Y. If the male contributes a Y chromosome, it combines with the female X chromosome to result in male offspring. If the male donates an X chromosome, the offspring is female. The sexual determination of offspring is decided when both parental chromosomes combine in the first nucleus of the ovum (Fig. 1-3).

Dominant and recessive inheritance

At the molecular level the interactions of the structural genes and the regulator genes influence the modifications of individual cell morphology. At the general level the phenotype, or physical characteristics of a person, is the representation of the genotype, or genetic traits of that person. Expression of the genotype depends on the difference or sameness in the two alleles at corresponding points in two homologous chromosomes. If the two alleles are different (one representing a dominant trait and the other a recessive trait), the person is heterozygous for that trait. If the two alleles are identical (being both dominant or recessive), the person is homozygous for that trait.

In heterozygous persons the dominant trait is expressed, whereas the other trait remains recessive and unexpressed. In homozygous dominant expression, both alleles carry the same trait, as they do when a homozygous recessive trait is expressed. However, the distinction of a dominant homozygous from a dominant heterozygous expression by pure clinical observation of that person is often impossible.

A complete discussion of the disorders inherited through the autosomal recessive genes is continued in Chapter 6.

Genetic linkage

Another factor influencing genetic outcome is linkage. In this instance, physically close genes in the same chromosome tend to be inherited together. Thus they are referred to as linked genes. Much research is necessary before positive predictions based on disease-associated traits will be possible. Little is known about linkage currently, except in the case of sex-linked inheritance.

Hemophilia, the classic example of this mechanism, is a sex-linked disorder that is most commonly transmitted by affected females and expressed in males. Females who transmit hemophilia to their sons must be heterozygous for the trait. Should a hemophiliac male mate with a homozygous (normal) female, the sons will not express the disease, but all of the daughters will be heterozygous or carriers of the trait.

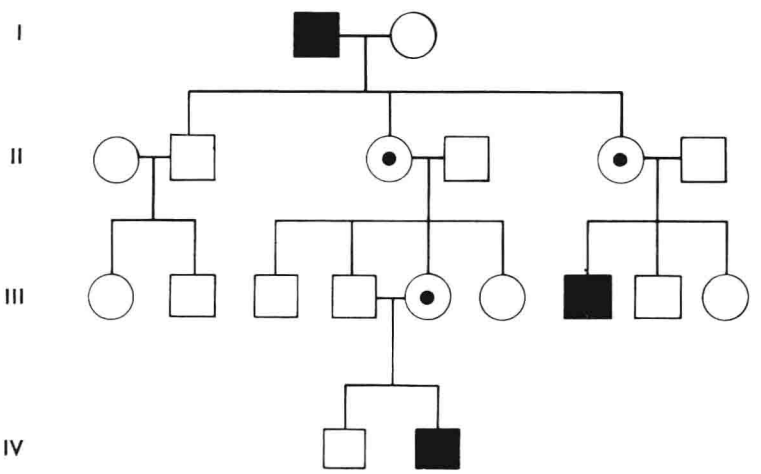

Fig. 1-4. Pedigree showing typical sex-linked recessive inheritance. Females who are carriers are indicated by a dot within circle; males who are affected are indicated by blackened squares. (From Chinn, P. L.: Child health maintenance, St. Louis, 1974, The C. V. Mosby Co.)

Therefore the disease will not be evident in that generation, but will be expressed in half of the male offspring produced by the affected (heterozygous) females (Fig. 1-4).

Variable expressivity

Another poorly understood phenomenon is that of variable expressivity. In this situation, persons inheriting given genetic traits have these in differing degrees. One person may have a very prominent trait, whereas another person, even within the same family, may have a mild expression of the trait.[6]

EMBRYONIC DISC FORMATION

After the second maturational division is completed the actual cellular development of the zygote begins with its division into two cells called blastomeres. By the end of the third day the blastomeres have increased to a 12-cell mass shaped like a mulberry and known as the morula. Eleven of the cells in the morula surround a larger central cell that later becomes the embryo. Peripheral cells form the protective membranes for the embryo.

In subsequent divisions the embryonic cell retains the greatest number of formative cells, divides slowly, and contains a large amount of the fertilized ovum. In contrast, the supportive cells undergo rapid multiplicative growth, at the expense of size and potency, to become trophoblastic cells.

By the fifth day the blastocyst, a free-floating intrauterine mass of 60 cells, has formed. It is composed of five formative cells, an increased intracellular volume, and trophoblastic cells at the peripheral edges. The external appearance has changed from the two-blastomere form into a single cavity. The formative cells of the blastocyst undergo multiplicative growth until they appear to become an irregular mass that attaches itself to the exterior wall of the uterus (Fig. 1-5).

Trophoblastic cells differentiate into two types of cells: polar trophoblasts and endoblasts. The polar trophoblasts assume a position at the periphery of the formative mass and soon provide an external protective cover. Other polyhedral trophoblasts, called endoblasts, provide the inner lining of the formative mass. This lining is the embryonic endoderm (Fig. 1-6).

Late in the sixth day the polar trophoblasts experience many changes that include rapid multiplicative growth and cell fusion that oblit-

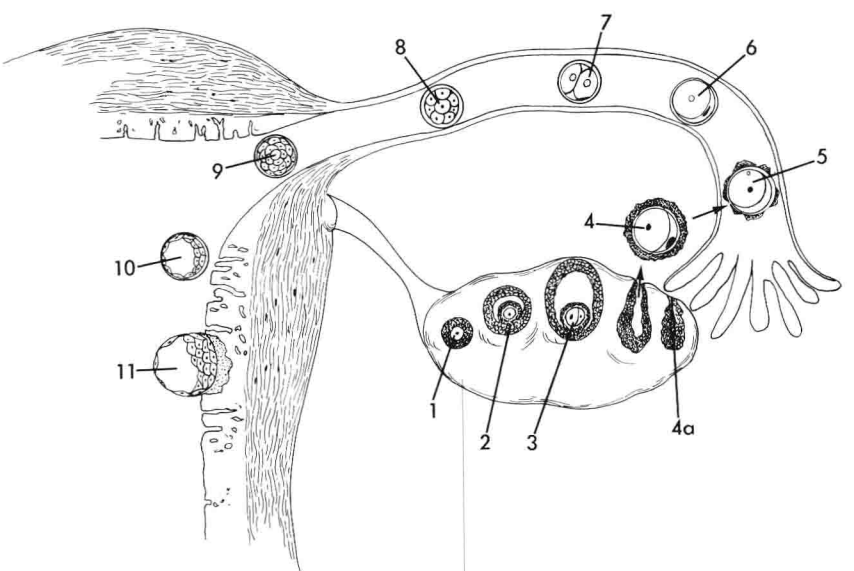

Fig. 1-5. Sequence of events leading to implantation. *1* and *2*, Early stages; *3*, formation of polar cell; *4*, mature ovum; *4a*, corpus luteum (early stage); *5*, fertilization; *6*, segmentation of nucleus; *7*, two-cell stage; *8*, eight-cell stage; *9*, morula; *10*, blastocyst free in uterine cavity; *11*, implantation.

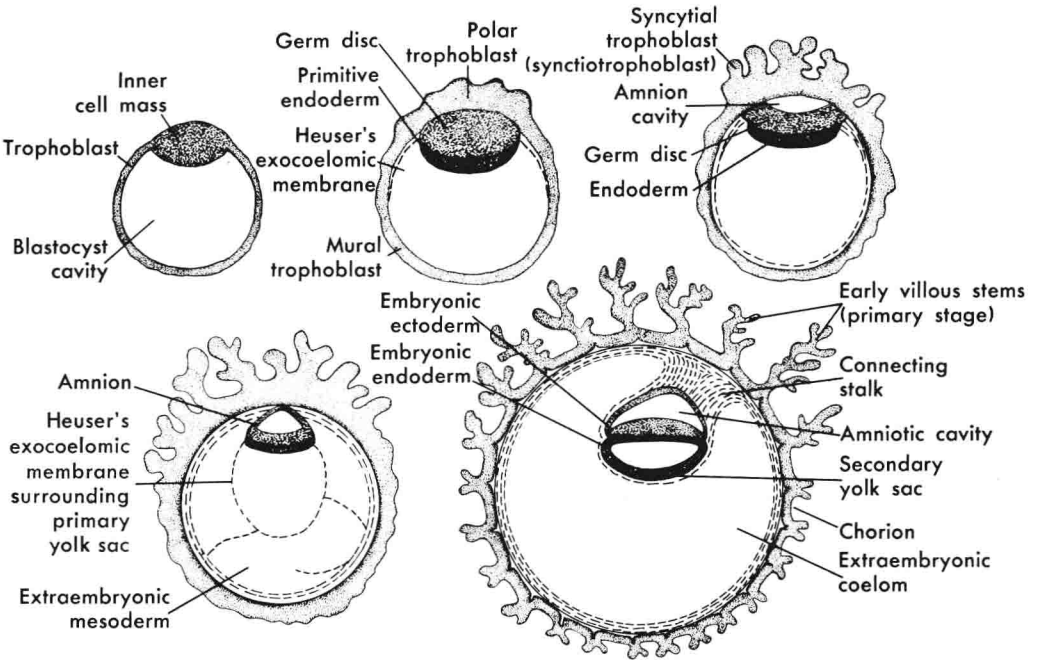

Fig. 1-6. Early stages of cellular differentiation.

erates individual cell identity as a result of the histolytic activity present as the polar trophoblast affixes itself to the uterine mucosa and engulfs degenerating uterine tissue. Within 9½ days this process has enabled the blastocyst to become deeply embedded in uterine mucosa, the area of deepest burrowing being the embryonic pole. The resulting penetration, growth, and fusion cause the polar covering to change into a multinucleated mass of cytoplasm called the syncytiotrophoblast. Subsequently, other areas of the blastocystic wall undergo changes, as the wall divides into an inner layer, the cytotrophoblast, and an outer layer, the synctial trophoblast (syncytiotrophoblast) (Fig. 1-6).

At this same stage the formative mass is composed of large, irregular cells in an external layer known as the germ disc, or embryonic ectoderm, and an internal mesodermal layer that separates this disc from the blastocystic cavity. At the periphery of this embryonic ectoderm, development continues that will ultimately divide this layer from the trophoblast and incorporate it into the amniotic cavity.

At the inferior end of the amnion an extension of possibly embryonic endodermal or cytotrophoblastic origin encapsulates the primary yolk sac. This membrane is known as extraembryonic endoderm or exocoelomic (Heuser's) membrane. The yolk sac is an essential source of sustenance for the growing embryonic ectoderm's continuing development (Fig. 1-6).

At this point the previously irregular embryonic ectoderm develops into columnar cells, which are bounded superiorly by a layer of amniogenic cells derived from the cytotrophoblast and inferiorly by the exocoelomic membrane that separates them from the primary yolk sac.

New avenues for sustenance are provided for the growing amnion as the penetrating synctium invades the nutrient-rich maternal blood vessels. These same pathways become the precursors of the placenta (Fig. 1-6).

Next the columnar epithelium of the embryonic ectoderm forms the ectodermal disc, a distinct entity of the amnion separated by a basement membrane from the endodermal cellular layer.

During the elapsed time between the development of the trophoblast and the amnion, delaminating mesodermal cells from the cytotrophoblast and some coagulum form loose re-