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CHAPTER 1

Introduction

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Why there is a need for this book

Pediatric, adolescent, and young adult gynecology is not taught because of our traditional compartmentalized system of medical school education. Our gynecologic residents never see children and would be ill equipped to evaluate a pediatric gynecologic problem. Our pediatric house staff rarely learn about gynecologic problems or how to evaluate those problems in their young patients. There are only a handful of fellowships in the USA in pediatric and adolescent gynecology. Nevertheless, it is important for the clinical physician to have a basic, practical understanding of diagnosis and management of common problems and to know when to suspect a possibly dangerous situation.

In the future, medical school curricula will develop a multidisciplinary approach and will include this subject. The specialty examinations for obstetrics/gynecology and pediatrics in the USA now have questions on the subject. This enables the student to immediately understand the relevance of a preclinical science such as gynecologic pathology. In our school it is now taught with clinical case presentation, physical examination, ultrasound diagnosis, and management.

The child is completely different from the adult in every aspect including approach, evaluation, and management. Their illnesses are different, such as tumors, problems of growth and development, future fertility, and delayed and precocious puberty. As an example, whereas the first procedure for abnormal bleeding in the menopause is usually endometrial sampling or dilation and curettage of the

uterus, in the adolescent this procedure is the last choice. The reason is that menopausal bleeding may be due to endometrial cancer which is extremely rare in the adolescent. In addition, in the latter it may result in emotional and possibly local physical damage.

Adolescent ovarian malignancies are usually of the germ cell variety. These have a different biology from the usual adult ovarian surface serous papillary cancer. The latter immediately spreads to the opposite ovary and is usually detected in an advanced stage. Chemotherapy often appears successful but after a negative second look there is often a recurrence. The germ cell malignant tumor of the child rarely spreads to the opposite ovary. Without treatment, it is rapidly fatal. However, with removal of the involved ovary and adjacent tube, resection of metastatic lesions and with current chemotherapy (different from that for the adult), most cases can be cured and fertility preserved.

In former years the rare vaginal rhabdomyosarcoma was usually fatal within two years unless it was discovered early and a pelvic exenteration performed. At present, with early diagnosis, local resection and chemotherapy, most cases are cured.

Unfortunately, humans develop sexual and reproductive ability in adolescence before achieving an adult sense of responsibility, completing basic education, and achieving financial independence. Despite education, sexual activity occurs and with it come problems of sexually transmitted infection and emotional stress. The USA has a high rate of unwanted pregnancy because of a lack of contraception information and access. Pregnancy complications may present unexpectedly such as ectopic pregnancy, miscarriage, and hydatidiform mole.

The adolescent may be exposed to substance abuse (smoking, alcohol, drugs), being overweight, eating disturbances (anorexia nervosa, bulimia) and automobile

accidents (when allowed to drive). She often fears that she is not developing normally, and struggles to be independent. Although not common, adolescent suicide is of concern. The US Centers for Disease Control and Prevention reported for 2003–2004 that the suicide rate for ages 10–24 increased by 8%, the largest rise in 15 years, to 4599 or 7.32 per 200,000. The increase was especially high in female adolescents. There are differences of opinion over whether this was a statistical fluctuation or the start of a trend, whether the reduction in prescriptions for antidepressants was a factor, and whether it was related to increased use of alcohol or drugs. A similar suicide increase also occurred in The Netherlands [1].

In the standard adult gynecology practice, many patients can be seen in a relatively short time because of the ease of history taking, examination, evaluation and explanation to the patient. Dealing with the young patient takes more time in obtaining a history from the patient (if old enough) and/or parents, and requires a quiet, unhurried individualized, nonintimidating approach and a sense of privacy for the apprehensive adolescent. The physician should be aware of legal aspects and cultural and family traditions. Nevertheless, there comes a time when complete privacy of the adolescent should be maintained, even from the parents. This is necessary because otherwise the adolescent may not report dangerous activities. The adolescent has to develop a personal sense of individual responsibility for her well-being. There should be separate clinic times for young patients and ideally the staff should have empathy for and like young patients.

When the adolescent is covered by the parent's health insurance, this may raise concern about the secrecy of health records and tests.

Despite the difficulties of caring for the young patient, the provider will derive great personal satisfaction helping youngsters have a happy, healthy, motivated, productive life with high self-esteem. This will stimulate their seeking higher education, assuming leadership roles if desired, and avoiding unwanted pregnancy.

Thus the roles of medical care include education, prevention of illness, treatment, and promoting good physical and emotional health.

Why this book is different

This book is a current, concise, clinical, practical, reader-friendly overview and guide for common problems. It is

directed to the practicing obstetrician, gynecologist, pediatrician, adolescent medicine physician, and concerned primary care physician. It emphasizes diagnosis and management. Clinical chapters are structured to give an immediate overview for an urgent problem and further details for when more time is available. Throughout the book there are Quick Take boxes that list critical points. Thus, when the apprehensive mother and patient burst into your consultation room screaming "She is hemorrhaging! Do something!" you will not panic.

Recommendations are given regarding referral for complex, rare, urgent or possible serious problems.

For many years I have conducted an annual postgraduate course with an ever-increasing audience on "pediatric and adolescent gynecology." Recently I have added the "young adult." Many gynecologic problems which begin in the adolescent continue into young adulthood. These include menstrual disturbances, dysmenorrhea, pelvic pain, polycystic ovarian syndrome, anovulation, obesity, eating disturbances, and early diabetes. With sexuality, there are problems of unwanted pregnancy, disruption of education, sexually transmitted disease (STD), papilloma virus (HPV), and immunodeficiency virus (HIV, AIDS). There are also the emotional stresses of puberty such as depression and the temptation of substance abuse (smoking, alcohol, drugs) and gambling. In the USA, there is significant mortality and morbidity from automobile accidents. Furthermore, there is no uniform agreement on the boundary between puberty and young adult. There are many criteria including chronologic age, menstrual age, bone age, sexual development, emotional maturity, etc. Different populations and individuals in each population may vary in maturation. Different cultures have different definitions of adulthood. Young adulthood as a topic itself has been overlooked and in addition the young adult tends to avoid routine care.

This book is intensely personal, based on many years of solo private practice in the same office in New York City (Manhattan), with continuous voluntary academic affiliation at Mount Sinai Hospital and with additional affiliation to help the initiation of the new Albert Einstein College of Medicine in the Bronx, New York City. In addition, I conducted three pediatric gynecology clinics at three hospitals. The one at Lenox Hill Hospital was held weekly for a continuous 20 years. My large private consultation practice continues where the same patients are seen continuously over many years.

Thus this book is the first to describe new observations, including the following.

- The prepubertal unilateral, rapidly growing enlargement of the labium majus without a border simulating a malignancy: “prepubertal unilateral fibrous hyperplasia of the labium majus, report of eight cases and review of the literature” (Chapter 25) [2].
- The prepubertal distal longitudinal vaginal folds which may be confused with rhabdomyosarcoma and sexual molestation (Chapter 26) [3].
- The first description of subacute atopic dermatitis of the vulva which may be confused with sexual molestation. Dermatologists erroneously think it rarely involves the vulva. Atopic dermatitis is increasing in the USA and industrial areas (Chapter 7) [4].
- The first reported case of xanthogranuloma of the vagina which, despite benign histology, requires close follow-up (Chapter 8) [5].
- The first report of using human cultured bilayered newborn foreskin to line the vaginal dissection in Rokitansky syndrome (congenital absence of the vagina and uterus in a young adult). It had been thought that laboratory-cultured skin would lose its antigenicity and not be rejected by another human. Immediate early rejection did not occur but late slow rejection occurred two weeks later. Nevertheless, there was a good long-term result due to the extraordinary multiple growth stimulation factors which speeded the inward ascent of histologic vaginal cells from the 2 cm patch of the vaginal dimple (remnant of the embryologic urogenital sinus) (Chapter 24) [6].
- Successful *in vitro* culture of human skin and vaginal mucosa from the vaginal dimple and vagina in a three-dimensional, relatively thick layer of bovine collagen. This should withstand the pressure of the soft vaginal surgical mold and later use of a firm stent for the Rokitansky syndrome (and other reconstructive surgery) and be transplanted to the original donor without rejection. It would be easier to handle and less susceptible to trauma than the usual delicate epidermal skin culture for use in burn victims which cannot tolerate pressure and which often heals with scarring (Chapter 24) [7].

Because of wide experience of about 100 cases of Rokitansky syndrome, treated mainly by the modified McIndoe technique, there is a chapter describing in detail the psychologic, surgical, and follow-up care. A small error at any stage can cause a poor result (Chapter 24).

Evolving practice concepts

Concepts and practice are constantly changing and evolving. Laparoscopy has a role for benign ovarian cystectomy and is also beginning to be utilized in malignancy (Chapters 20, 26, 37). Pelvic and renal ultrasound may expand to include perineal and transrectal approaches. Magnetic resonance imaging (MRI) is being used especially for anomalies. Serum tumor markers are increasingly utilized. There is a fetal protective approach to congenital adrenal hyperplasia (CAH). The more common, atypical, late-onset CAH is being appreciated as a factor in infertility, polycystic ovarian syndrome, acne, hirsutism and scalp balding of the adult (Chapter 4).

Herpes simplex type 2 is increasing in incidence. Most cases are asymptomatic but nevertheless may shed virus and be infective to their consort. Management of pregnant women now includes herpes simplex type 2 antibody status. If negative and the partner is positive, he may be shedding virus and should consider taking medication to prevent spread. If the patient is positive, should she take suppressive therapy at the end of pregnancy to avoid a vulval ulcer in labor and a cesarean section (Chapter 38)?

The American College of Obstetricians and Gynecologists (ACOG) recommends that the first visit to the obstetrician-gynecologist should be between the ages of 13 and 15 years with the mother present. The visit is the start of a physician-patient relationship with a discussion of preventive healthcare, education and guidance, and any specific individual concerns of the patient. At the initial visit usually there is no physical or pelvic examination unless indicated. A simple approach might be a concise introduction followed by referral to group educational talks by nurse practitioners and/or appropriate books or educational videos. If feasible, try to schedule repeat visits perhaps at six-monthly intervals without interrupting school, and when other patients of a similar age group are being seen. The patient and her parents should be aware that regarding confidentiality, the law requires that the risk of bodily harm and physical or sexual abuse of minors is reported. Educational topics might include puberty development, menstruation, menstrual diary, dysmenorrhea, prevention of pregnancy, condoms, emergency contraception, prevention of STD, and avoiding substance abuse, eating problems, and mental health problems (anxiety, depression). A preinterview questionnaire as well as visual teaching aids are helpful. A visual general body evaluation,

including visual breast evaluation and axillary and pelvic hair with perineum visualization, might be done at the initial or later visit to evaluate development, depending on patient anxiety and medical indication. If the patient is sexually active, she should be screened for STD including new urine STD screening if needed to avoid the need for a vaginal speculum.

The ACOG recommends that the first Pap cervix cytology and HPV cervix test be delayed until three years after the first coitus (or by age 21) because the first HPV infection may cause an over-reported cytology which may provoke surgical procedures which might disturb future pregnancy and because by three years most HPV tests become negative (see Chapters 38, 40, 45). However, other tests may be performed when there is sexual activity, including gonorrhea, chlamydia, etc. [8].

A problem in the USA is the multiple insurance companies which administer health maintenance organizations and managed care. There may be unavoidable physician changes as the parents alter their insurance companies with employment changes. This may mean a new physician introduction with associated stresses. Despite good intentions, adolescents are often lost to medical care.

Another problem is that a significant proportion of the population does not have health insurance.

Vaccination for human papilloma virus

In addition to preserving good health, vaccinations is a method of encouraging continuity of routine care [9]. It gives the opportunity for guidance in avoiding risk-taking behavior and substance abuse, especially for adolescents with poor self-esteem, depression, stress, and inadequate social support [10].

The high-risk type of papilloma virus, hrHPV (of which there are many), is a sexually transmitted disease. Although most sexually active young people acquire HPV, usually without symptoms, most spontaneously cure themselves within three years. In about 10% it may persist and may cause cervix dysplasia and later cervix cancer. In the USA there is an approved vaccine which will prevent most types of high-risk virus and is recommended for girls prior to sexual activity. It has not as yet been approved for boys. It is highly recommended by the medical profession to reduce the chance of cervix cancer by about

75% [11, 12]. The vaccine requires three separate injections and is expensive. Vaccination is approved for ages 9–26 regardless of previous sexual activity (Chapter 40).

The trade name of the quadrivalent HPV type 6, 11, 16, 18 recombinant vaccine is Gardasil[®]. The US Food and Drug Administration approved it for females aged 9–26 years in 2006. The Advisory Committee on Immunization Practices recommended vaccination at ages 11 or 12. The ideal situation is vaccination before sexual activity. The original recommendation of the manufacturer (Merck & Co., Inc.) was that vaccination would not do any good once there is a positive hrHPV cervical cytology test. The vaccine is given in three doses at 0, two and six months by injection.

The current viewpoint is that vaccination is helpful despite previous sexual activity, cervix dysplasia, and positive hrHPV. The reason is that most cases of hrHPV infection are with one or, rarely, two genotypes of hrHPV. The standard hrHPV test does not specify which genotype is present and will give a positive result for any of about 15 hrHPV genotypes. Although less effective in preventing later possible cervix cancer, it is now thought appropriate to give the vaccination despite a positive hrHPV test.

About 15 hrHPV genotypes are associated with cervical cancer. About 70% of cervical cancer is associated with hrHPV genotypes 16 and 18. About 90% of genital warts (which are usually not malignant) are associated with HPV genotypes 6 and 11.

In 2008 a vaccine was approved to prevent vaginal and vulvar cancer. Despite vaccination, standard cytology screening should be continued, starting three years after first coitus in the adolescent or no later than age 21. Routine cytology and HPV screening should be performed at age 30 [13]. If hrHPV is positive at this stage, the patient must be carefully followed because cervical cancer may result from persistent hrHPV. Cytology is not always reliable so even a report of atypical squamous cells of unknown significance (ASCUS) merits consideration of colposcopy and biopsy. Some advise colposcopy and biopsy even with negative cytology. In addition, consider simultaneous endocervical curettage, especially for atypical glandular cells on cytology which could indicate primary endocervical glandular neoplasia which is increasing in young women. It tends to be discovered late because it is not palpable, often has false-negative cytology, is not visible to direct view on ultrasound, and has a relatively poor prognosis.

Another concern is sexuality in later years. About half of marriages end in divorce in the USA. Widows and single women over 30 may have new sexual exposure and with it sexually transmitted infection. Although the FDA approval for HPV vaccination is up to age 26, this is simply because the over-26 year age group has not been sufficiently studied. Logically this group should consider vaccination.

For children, the physician should emphasize that vaccination is considered a standard recommendation used to prevent cervical cancer. It does not promote promiscuity.

HPV vaccination is not FDA recommended for pregnant women.

Most vaccinations are given to those with insurance coverage. Unfortunately, in the USA a significant proportion do not have medical insurance. However, with extensive use of cervical cytology (Papanicolaou) testing, cervical cancer rates and deaths have been greatly reduced in the USA.

In underdeveloped countries with poverty and poor-quality medical care, there is a lack of screening for cervical cytology and a much higher rate of cervical cancer and deaths than in the affluent countries. These countries would benefit even more than the affluent countries if HPV vaccination became available to them.

The Centers for Disease Control (CDC) in the USA now emphasize vaccination for both HPV and meningococcus for adolescents by age 18 [12, 13].

The adolescent or young adult patient should also consider vaccination or update for the following: hepatitis B and A, diphtheria, tetanus, pertussis, measles, mumps, rubella, varicella, meningococcus, pneumococcal, yearly influenza, (haemophilus influenzae type b), meningitis, and inactivated polio.

Education for life

As part of normal customary schooling, the young patient should be instructed in “education for life,” emphasizing good general physical and emotional health including exercise, diet, and avoiding obesity and substance abuse. In addition, she should understand the importance of good education, self-esteem, empowerment and having aspirations for adult life, including the later choice of a mate. Learning human biology is important. Because sexual development occurs before adulthood, the young patient requires early guidance.

A recently recognized concern is universal increasing obesity in developed countries which often starts in adolescence. It results from a combination of a genetic tendency to “truncal obesity” (waist-to-hip ratio), intolerance to the refined carbohydrates of affluent societies, and lack of exercise. This may affect up to 30% of all humans. Obesity predisposes to diabetes and cardiovascular disease. The lack of exercise compounds the problem [14].

A school course on contraception is often met with opposition. If it is presented as part of an overall program on “education for life,” it is more acceptable. Parents and school administrators sometimes erroneously believe that it may encourage promiscuity. The USA has a high rate of adolescent unwanted pregnancy. Other developed countries have similar rates of adolescent sexuality but without unwanted pregnancy because of the use of contraception. Correspondingly we have high rates of abortion. Unwanted pregnancy is an important reason for school drop-outs as well as emotional stress. Teaching abstinence alone usually is ineffective.

The educator should not be judgmental and yet not condone sexuality. The young girl should be advised that she controls her own body and is empowered to do whatever she wishes, and that is not to have sex if she doesn't wish to. She can always say “I'm not ready for it yet.” If she is sexually active she needs contraceptive advice.

Even though one hears that “everyone is doing it,” early sexual activity has hazards. First, there is the possibility of unwanted pregnancy. Even with oral contraceptives, there is a difference between the small theoretical risks of pregnancy compared to the actual risk in use, which is much higher because of forgetfulness. An unwanted pregnancy is usually a high-risk pregnancy associated with prematurity, disruption of education, substance abuse, and persistence of poverty.

Second, there is the possibility of sexually transmitted disease (STD) also called sexually transmitted infection (STI). STD may leave a legacy of infertility, ectopic pregnancy, pelvic pain, and AIDS. Condoms will tend to reduce the risk and therefore should be used in addition to more effective contraception. In addition, the number of consorts should be kept to a minimum. Routine testing for STD should be performed even in the absence of symptoms. Females acquire the HIV (AIDS) virus more readily than males because of a large susceptibility area of vaginal mucosa which frequently has microabrasions.

Even if the boyfriend is in apparently good health, he may be a carrier of STD.

The theoretical ideal would be to have the consort tested for all STD (STI) (Chapters 38, 39, 40). Early AIDS is asymptomatic. About one-third of the population in the USA have herpes simplex type 2 infection. Most are asymptomatic but can still shed virus and be infective. Syphilis and hepatitis are also of concern. Unfortunately although in many parts of the USA in previous years a blood test for syphilis was required for a marriage license, at present many communities do not require any testing (Chapter 44).

From a practical point of view the potential partner should be asked about previous number of partners, any past history or symptoms of STD (STI), drug addiction, sharing of intravenous needles, and hemophilia with many blood transfusions before complete blood testing was available.

From a legal viewpoint there have been law suits against adults for failing to report a history of herpes simplex type 2 to the partner who later developed it.

Personal observations

Dermatologic lesions of the vulva may be readily misinterpreted as molestation, especially with lichen sclerosus, atopic dermatitis, and distal longitudinal vaginal folds (Chapters 7, 34, 35).

Recurrent urinary tract infection may be due to a urogenital sinus, congenital adrenal hyperplasia (CAH), labial fusion, agglutination of the labia minora or microperforate hymen with the opening near or under the urethra. It may also occur from increased anal bacteria colonization in the vestibule and periurethral area in atopic dermatitis. Any vulvitis may cause vulvar dysuria with infrequent voiding and "holding it in." Voiding in warm water may be helpful.

Recurrent prepubertal vaginitis may be due to atopic dermatitis with perianal fissures allowing anal bacteria to reach the anestrogonic vagina. It may also result from rubbing the vulva because of pruritus. Recurrent atopic dermatitis may occur from secondary bacterial infection. Other causes include recurrent foreign bodies, congenital vaginal anomalies, and wearing of tights.

Recurrent and severe labia minora agglutination is often due to atopic dermatitis. Topical estrogen cream once

daily at night usually will cause spontaneous separation of the labia minora within two weeks. If the child requires diapers then the cream is removed when the child is cleaned. Therefore the cream should be applied three times daily. If the cream is used for several months it may be absorbed and cause breast stimulation, pain and tenderness, darkening of the vulva skin and vulva hair growth. If the labia minora have been agglutinated for about six months or longer, fibrous tissue may grow in and surgery may be required. It is best done by someone with experience because the urethra is just under the closed labia minora, especially if the labia appear to be completely closed. It may cause urinary tract infections, distortion of the voided urine stream, and postvoid dribbling. On examination there seems to be no exit for urine. With careful inspection, the most anterior section will be found to have a 1–2 mm opening with a watery appearance.

Agglutination which is less than half with an anterior opening not distorting the urinary stream usually does not require surgery.

In former years some physicians would rip apart the labia with both thumbs. This causes pain, bleeding, and a very disturbed patient and mother. In addition, the new jagged edges promptly reattach.

Some physicians recommend avoiding any treatment in anticipation of pubertal estrogen opening the labia. In this case, the latter remain closed for years and do not open at puberty because of agglutination being replaced by fibrous tissue.

For mild cases of labia minora agglutination, try good hygiene and topical petroleum jelly (Vaseline®).

House physicians never see children with agglutination of the labia minora because they are not hospitalized. The differential diagnosis includes CAH. Sometimes with atopic dermatitis, the foreskin traps smegma under the clitoris, causing severe local pain, or sometimes it causes swelling of the clitoris prepuce foreskin, simulating a large clitoris. With labial agglutination, this may simulate congenital adrenal hyperplasia.

Premature thelarche (prepubertal isolated nonprogressive breast development) is more common than true precocious puberty. Breast enlargement is seen in both. Although many experts recommend investigation with sometimes frightening x-ray of the left wrist for bone age, and extensive (and expensive) serum hormone tests, the latter may be difficult to interpret because of lack of biologic activity of elevated gonadotropin which is not

infrequent in the young child. I have found that most cases can be managed by examination, vaginal swab maturation index (or using centrifuged voided urine if not feasible), and periodic observation. Pelvic ultrasound may be performed to rule out an estrogen-secreting ovarian neoplasm (Chapters 43, 49, 52). Severe hypothyroidism may precipitate an increase of prolactin which may stimulate the breasts. The vaginal mucosa is more sensitive to estrogen than the breast. In the usual case of premature thelarche, the vaginal mucosa is anestrogenic, prepubertal, and thin and the cytology shows predominantly parabasal cells which are small, oval and contain a large nucleus.

With true central nervous system precocious puberty there is increase in serum gonadotropin and estradiol, and the vaginal mucosa is thick and has superficial, large, flat polygonal epithelial cells with a small dark nucleus. The vaginal mucosa changes before the breasts enlarge. The educated observer can confirm this immediately by a wet microscopic swab without staining. With an atrophic smear there is no estrogen from true central nervous system activation or rare ovarian neoplasm. With premature thelarche both breasts may enlarge simultaneously or one may enlarge followed by the other in a few months. The breast enlargement ceases spontaneously and does not get larger than stage 3. It is thought that premature thelarche may result from a previous transient elevation of active gonadotropin. Therefore there are rare in-between cases (Chapters 18, 27).

Old autopsy reports of rhabdomyosarcoma show that recurrences always present at the original site. This suggests that the site should be tested after therapy. I performed a local vaginal resection of a lesion high on the anterior wall in an 11-month-old girl. A simultaneous deep bladder biopsy showed the tumor in the vesicovaginal septum. After appropriate chemotherapy, it was thought that the tumor was cured. However, repeated vaginoscopy showed recurrences at the site which had repeated excisions. For the next year there were repeated courses of chemotherapy. The local recurrences persisted and after one year radiation and anterior exenteration had to be undertaken. She has remained tumor free after 20 years (Chapters 8, 37, 49).

Almost every gynecologic problem in the young patient has a psychological component (Chapter 13).

If surgery can be accomplished without excessive delay following torsion of the ovary or adnexa, despite an ominous dark blue color and swelling, detorsion plus careful

observation for the next 5–10 minutes will often show that circulation is restored or improved and the structures can be preserved. A predisposing dermoid cyst should be resected to prevent recurrence of torsion. There are differences of opinion regarding pexy of one or both ovaries (Chapters 26, 32, 33, 36).

Cervical histology of the adolescent shows the dramatic, initial HPV infection. The marked reaction explains the often alarming cytology which may lead to overly aggressive surgery (Chapter 45). Initial viral infection sites tend to have violent reactions as the host mounts a vigorous immune response. This is analogous to a primary genital type 2 herpes simplex virus infection. Later biopsies are subdued. The current concept is to defer cytology to about 2–3 years after the first coitus by which time, in the vast majority, the cytology returns to normal and the DNA test for HPV becomes negative by an unknown normal body defense mechanism. Nevertheless, tests for other STD should be performed with the first coitus and repeated periodically, as well as education regarding contraception, pregnancy complications, and AIDS (Chapters 38, 39, 40).

Sometimes, often after a broken partnership, if the asymptomatic patient requests tests for all STD, consider doing cervical cultures or DNA tests for gonorrhea and chlamydia, serum tests for syphilis, HIV (AIDS), herpes simplex type 2 specific antibody, hepatitis screen, lymphogranuloma venereum or any other STD suggested by cultural or geographic factors. In addition, consider inspection of the throat and skin and palpation for inguinal adenopathy (Chapters 39, 40).

The use of ultrasound (sonogram) may reveal unsuspected fetal ovarian cysts which often persist into the early months of infancy. Most of these are benign follicular cysts which are reminiscent of the approximately 10% incidence of postmenopausal asymptomatic, benign, small simple ovarian cysts which may come and go and are discovered by coincidental transvaginal ultrasound. It is not unusual for follicular cysts to develop in the prepubertal child. These are usually asymptomatic and tend to disappear within six weeks to a few months (Chapters 9, 36, 50).

Often coincidental pelvic ultrasound may reveal a “complex ovarian cyst” in an adolescent or young adult. These are usually benign asymptomatic hemorrhagic corpus luteum cysts which usually recede in six weeks. Some are associated with early pregnancy. In the

postmenopausal woman, a “complex ovarian cyst” should arouse concern (Chapter 36, 41).

Although polycystic ovarian syndrome (POS) is associated with oligomenorrhea and amenorrhea in the young adult, I have noticed that in the adolescent it may manifest by irregular, heavy vaginal bleeding. This is understandable because anovulatory dysfunctional uterine bleeding (ADUB) may manifest by irregular heavy bleeding or, less often, by a characteristic three months of amenorrhea followed by one month of continuous irregular bleeding. About 5% or more cases of ADUB, especially those with heavy irregular bleeding, may never ovulate and may later be diagnosed as POS. About 5–25% of POS may be due to atypical late-onset CAH (Chapters 23, 32). Apart from immediate care, POS has implications for later general health and therefore for preventive care.

Most vaginal bleeding in the child is due to local causes rather than precocious puberty. Even children with signs suggestive of puberty may have bleeding from a local cause (Chapter 8, 10, 11).

Chronic pelvic pain is not rare. Sometimes the etiology is uncertain and often it is attributed to a coincidental benign simple small unilocular ovarian cyst. Not infrequently, there is a gastrointestinal etiology suggested by bilateral continuous or intermittent crampy pain, tympanic abdomen on percussion and palpable right colon and sometimes sigmoid colon. It may be associated with lactose intolerance, food allergies, regional ileitis (spastic colon), irritable bowel syndrome, intestinal parasites, and celiac sprue. Sensitive skin stroking may suggest neurologic or fascial origin. Do not rush to perform a laparoscopy for recurrent pelvic pain if the patient has had several previous laparotomies or laparoscopies. There may be adhesions, nongynecologic cause of pain, and an increased chance of surgical complications.

Fellowship development

Co-ordinated advanced training is almost nonexistent in the USA outside a handful of fellowships, each of which is different. There is a need to develop a consensus standard fellowship program which would include two aspects: nonsurgical education, and surgical aspects for those with surgical privileges.

The main limiting factor for advanced training is the relatively few reconstructive procedures for congenital anomalies in one hospital done annually.

Personal concept of collaborative fellowship

Pediatric and adolescent gynecology, although a small subspecialty, is being recognized by all large hospitals and clinics as an important service. Each institution has different strengths for teaching programs. Surgical procedures are infrequent, especially for congenital anomalies. Thus it would take a relatively long time for a fellow to achieve sufficient experience at one hospital. Institutions are reluctant to close certain services in order to give complete care in one institution. The logical approach is to establish a collaborative fellowship in which the fellow will rotate through different institutions and in addition scrub in or observe surgery for unusual cases as they present themselves. In addition to training, such collaboration could facilitate research planning, data collection, and collaborative hospital educational conferences. At a certain stage, the fellow might even be a consultant for emergency rooms.

Since most hospitals are territorial in philosophy in an attempt to give full service, the ideal would be a collaborative rotating fellowship with one base co-ordinating hospital. This is especially true for New York City with its five medical schools and numerous competing “children’s hospitals.”

Central Europe has had a long traditional interest in advanced training (Chapter 52). There is a standard test and certification which could be used as the basis for a curriculum, test, and certification in the US.

Concerns associated with particular age groups

Fetus

Occasionally, by coincidence, a large simple follicular cyst of the ovary is discovered by routine ultrasound in pregnancy. The usual management is periodic observation by repeat ultrasound for the remainder of the pregnancy and after birth. Most cysts are asymptomatic and spontaneously disappear in a few months after birth. Infrequently surgery is performed in the newborn or infancy if there is evidence of malignancy or torsion (Chapter 9).

Newborn to about two weeks

- Atypical-appearing genitalia (ambiguous external genitalia) (Chapter 3)

- Congenital adrenal hyperplasia, salt-losing form may cause sudden death at one week (Chapter 4)
- Androgen insensitivity (Chapter 5). Family history of amenorrhea, infertility; bilateral inguinal hernia sometimes containing a testis; feminine vulva; short vagina; no cervix or uterus; karyotype 46,XY
- Ovarian dysplasia (gonadal dysgenesis, XO syndrome, Turner syndrome): karyotype 45,X mosaic; anomalies (cardiac, coarctation of aorta, renal, extremities), edema of the dorsum of the hands and feet; broad chest; excessive pigmented nevi (Chapter 5)
- Microperforate hymen (small orifice just below urethra) (Chapter 25)
- Imperforate hymen: sometimes bulging from excess endocervical mucus due to over-reaction to maternal estrogen (Chapter 25)
- Midvaginal complete transverse septum may result in a huge upper vaginal distension with cardiorespiratory distress (Chapter 25)
- Cyst of introitus (paraurethral cyst) present at birth concealing urethra and vagina, attached to distal anterior vagina and distal posterior urethral meatus (Chapter 25)
- No opening to vagina despite mucosal dimple – possible Rokitansky syndrome of absent vagina and uterus; test patency with adult urethral catheter (Chapter 24)
- Polyps of the hymen, usually small at 6 o'clock, may shrivel and disappear in a few days when maternal estrogen effect is lost. Some may persist and require removal for cosmetic reasons or bleeding from trauma (Chapter 25)
- Unusual vulva rashes due to secondary syphilis acquired in late pregnancy following early pregnancy negative serology (Chapter 7, 40)
- Associated anomalies – rectum, low back, spina bifida, renal anomalies, ectopic ureter (Chapter 25)
- Physiologic leukorrhea and bleeding due to maternal estrogen stimulation and later withdrawal bleeding
- Trichomonad vaginitis and urethritis acquired from maternal vagina after vaginal delivery can cause a heavy white discharge and sterile pyuria (Chapter 7)
- Labial edema due to breech presentation
- Rare vaginal rhabdomyosarcoma presenting as a polyp and/or bleeding with the base above the hymen. The surface may have normal vaginal mucosa with the neoplasm being submucosal; therefore the entire polyp must be removed rather than a surface mucosal biopsy (average age for rhabdomyosarcoma is about 11 months) (Chapters 8, 37, 49).

- Bilateral slight breast enlargement, 1 cm cystic-feeling breast tissue, sometimes with galactorrhea (“witch’s milk”). Do not massage breasts which may provoke infection. If surgery performed for abscess, do not excise breast bud (Chapter 27).

Infant (about 2 weeks to 2 years)

- Diaper rash (Chapters 7, 36, 37)
- Atopic dermatitis (Chapters 7, 36, 37)
- Seborrheic dermatitis (Chapters 7, 36, 37)
- Agglutination of labia minora, often associated with atopic dermatitis (Chapter 7)
- Vulvar and perianal condyloma: may be acquired from vaginal birth exposure to maternal vaginal papilloma virus and/or by caretaker cleansing, especially if there is sensitive skin. It may be sexually transmitted (Chapters 7, 38, 39, 40, 45)
- Premature thelarche, unilateral or bilateral (Chapters 18, 27; also see above p. 6)
- Many newborn concerns

Young child (about 3–6 years)

- Trauma (climbing, running, falling) (Chapter 10)
- Foreign body causing persistent, foul bloody discharge, usually due to wads of toilet tissue deep in the vagina (Chapter 7)
- Agglutination of labia minora (Chapter 7)
- Vulvovaginitis (Chapter 7)
- Atopic and seborrheic dermatitis (Chapter 7)
- Prolapsed urethra (Chapters 1, 8)
- Ovarian follicular cysts (usually asymptomatic), coincidental discovery (Chapters 9, 50)
- Premature adrenarche (pubic hair, may be familial) (Chapter 18, 27)
- True central nervous system precocious puberty, with ovulation, high follicle-stimulating hormone (FSH) and estradiol (Chapter 18)
- Rare rhabdomyosarcoma of vagina (Chapter 8, 37, 49)
- Rare hormone-producing ovarian cystic tumors (granulosa cell) causing precocious puberty without ovulation, elevated estrogen, and no increase in FSH (Chapters 18, 47)

Prepubertal (about 7–10 years)

- Vulvovaginitis (Chapter 7)
- Vaginal bleeding (foreign body, rare rhabdomyosarcoma) (Chapter 8)

- Trauma (Chapters 8, 10, 11)
- Ovarian follicular cysts: usually asymptomatic, discovered by coincidental ultrasound, done for nonspecific lower abdominal cramps. A simple, small unilateral cyst may be coincidental. Usually repeat ultrasound after about two months shows resolution (Chapters 9, 36, 50)
- Complex ovarian cysts (dermoid, hemorrhagic) may undergo torsion with adjacent tube (Chapters 9, 36, 50)
- Sexual molestation (Chapters 10, 11)
- Rare hormone-producing ovarian neoplasm (Chapters 41, 47)
- Ectopic ureter exiting into vagina causing constant watery purulent leakage (Chapters 7, 25)
- Beginning “education for life” courses, including puberty development and preparation for menses (see above)
- Rare unilateral rapid enlargement of labium majus (Chapter 25)

Puberty (about 10–17 years)

- Anxiety, emotional stresses
- “Am I developing normally?” (Chapters 13, 15, 27)
- Menstrual problems: irregularity, menorrhagia, dysmenorrhea, primary and secondary amenorrhea (Chapters 15–22, 24, 25)
- “Education for life”, including contraception, avoiding STD (sexually transmitted infection – STI), continuing education, self-esteem, finding a partner, career development, obesity, substance abuse, development of a sense of responsibility, vaccination update (Chapters 15, 28, 29; also see above)
- Onset of manifestation of POS (Chapter 31) sometimes with excessive bleeding in adolescence and secondary oligoamenorrhea in young adulthood
- Acute pelvic pain: torsion of ovarian cysts (associated with long ligaments and heavy cysts) with tube (Chapters 32, 33, 47), rupture of malignant cyst, ectopic pregnancy, pelvic inflammatory disease, acute appendicitis
- Pelvic and abdominal masses (Chapters 32, 33, 47)
- Chronic pelvic pain (Chapters 20, 21)
- Ovarian simple cysts, hemorrhagic cysts (hemorrhagic cysts may have the ultrasound appearance of “complex cyst” – repeat ultrasound in 6–8 weeks; “complex cyst” in menopause is of concern) (Chapter 36)
- Continuity of care, need for general health vaccinations which promotes continuity (see above)

- Consideration of HPV vaccinations (series of three injections) (Chapter 40; also see above)
- Ovarian neoplasms: dermoid cysts (mature benign teratoma), malignant germ cell tumors (rapid growth, large, pain) (Chapter 47)
- Understanding adolescent labor (Chapter 30)
- Confusing congenital anomaly symptoms: amenorrhea in a normal-appearing adolescent (Rokitansky syndrome; Chapter 24); severe dysmenorrhea not relieved by oral contraceptives (“no eponym” syndrome of uterus didelphys; Chapter 25)

Young adult (about 18–25 years)

- Continuation of adolescent concerns including: menstrual disturbances, sexuality, STD, avoiding unwanted pregnancy (Chapters 12, 15–23, 24, 25, 29, 30, 35, 40, 42, 47) as well as obesity and substance abuse
- Abortion (Chapters 29, 43, 44)
- Pregnancy complications: miscarriage, ectopic pregnancy, hydatidiform mole (Chapters 33, 48)
- Ovarian cysts and neoplasm including dermoid (mature teratoma); rare germ cell malignancies; adnexal torsion (Chapters 32, 35, 47, 48, 50)
- Asymptomatic “complex ovarian cysts” usually managed by repeat ultrasound in 6–8 weeks in proliferate phase (Chapter 36)
- Chronic and acute pelvic pain (Chapters 20, 21, 32, 33)
- Pelvic and abdominal masses (Chapters 32, 33, 47, 52)
- Understanding adolescent labor (Chapter 30)
- “Education for life” courses (see above)
- Finding a partner
- Advanced education
- Career development
- Developing a sense of responsibility
- Does she need vaccinations? (see above)
- Primary care doctor or gynecologist for continuing care and vaccination update (see above)

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Further reading

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