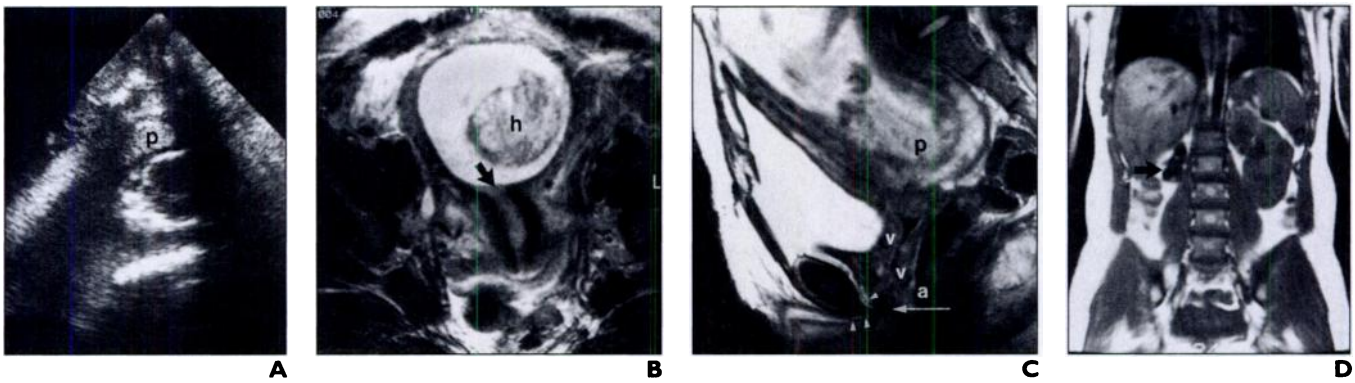

Fig. 4.—16-year-old girl with McKusick-Kaufman syndrome and term pregnancy.
A, Translabial obstetric sonogram (sagittal midline) shows inferior margin of placenta (p) extending to expected location of endocervix, but no cervical canal is seen. This finding was incorrectly presumed to represent marginal placenta previa.
B, Off-axis coronal fast spin-echo T2-weighted MR image (4000/108 [TR/TE_{eff}]) obtained with pelvic phased array coil shows endocervix (*arrow*) well away from placenta. Note fetal head (h), surrounded by bright amniotic fluid, above cervix. No evidence of placenta previa, increta, or percreta is seen. Placenta is cephalad and to right of endocervix, making placenta impossible to see in this imaging plane.
C, Midsagittal fast spin-echo T2-weighted MR image (4000/108) obtained with pelvic phased array coil of vaginal atresia after repair of neonatal hydrocolpos shows cloacal variant. Note placenta (p), thickened walls of upper two thirds of neovagina (v), air centrally recognized as signal void in atretic distal third of vaginal canal (*arrow*), and urogenital sinus (*arrowheads*)—that is, single common channel for urethra and inferior third of vaginal canal. Anal canal (a) appears anterior to normal location but separable from vagina and urethra.
D, Coronal abdominal spin-echo T1-weighted MR image (400/12 [TR/TE]) shows atrophic right renal remnant with hydronephroureterectasis (*large arrow*). Left kidney is normal. Findings are probably due to chronic reflux nephropathy with or without chronic atrophic pyelonephritis, which likely predated pregnancy. Extension of bladder (not shown) above level of umbilicus may have represented congenital form of megacystis.

Numerous cases of intramural air have been reported. The most common locations include the stomach [1], small bowel, and colon, and several mechanisms have been described. Infection with a gas-forming organism may dissociate all layers of the gastrointestinal wall and has been reported for the stomach [2]. Cystic pneumatosis often associated with chronic bronchitis or emphysema involves mainly the left colon and the stomach. The mechanisms for both the formation and the persistence of the cystic spaces remain controversial [3]. Interstitial emphysema may occur in the stomach [4], the small bowel, and the colon. Linear gas in the bowel wall suggests bowel infarction, although other conditions, including Crohn's disease, progressive systemic sclerosis, peptic ulceration, and trauma after gastrointestinal endoscopy, may be responsible for interstitial emphysema. The two basic underlying conditions are mucosal injury and increased intraluminal pressure. We hypothesize that in our patient, gastric dilatation was caused by premature cessation of nasogastric aspiration. The raised intragastric pressure led to the escape of air into the cardia wall through a mucosal tear caused by vomiting. Air dissecting along this pathway may continue superiorly into the esophageal wall and lead to retrosternal pain and nonspecific symptoms. The disappearance of intramural air could be explained by a decrease of intragastric pressure after nasogastric aspiration.

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Term Pregnancy in a Patient with McKusick-Kaufman Syndrome

The McKusick-Kaufman syndrome is a congenital autosomal recessive syndrome characterized by postaxial polydactyly and hydrocolpos [1, 2]. Modern approaches to the repair of the major cloacal malformations have reduced the high morbidity and mortality associated with the defects and have enabled the patients to pursue a more normal life. Here, we discuss a rare term gestation in a patient whose McKusick-Kaufman malformations had been repaired at birth.

A 16-year-old girl, gravida 1 para 0, presented to the obstetrics clinic at 18 weeks' gestation for prenatal care. McKusick-Kaufman syndrome with hydrocolpos and postaxial polydactyly had been diagnosed in the patient at birth. As a newborn, she had undergone surgical repair with dissection of her urethra and construction of a flap sutured to the distal vagina in an attempt to construct a separate orifice. She reported bouts of urinary incontinence and frequent urinary tract infections. Despite the inadequacy of this correction for intercourse, she had been sexually active since the age of 12 years, with only minimal penetration.

Upon examination of the patient, we noted a common urethral and perineal opening with the anus located proximally. Although no cervix was visualized, one was felt on rectal examination. Sonography revealed a normal intrauterine pregnancy of 18 weeks' gestation with a marginal placenta previa. The cervix was not seen sonographically. The bladder was determined to be anomalous, extending to the level of the umbilicus. Right renal agenesis was suspected (Fig. 4).

MR images obtained with a Signa 1.5-T scanner (General Electric Medical Systems, Milwaukee, WI) and a pelvic phased array coil showed the endocervical os; ruled out a placenta previa, increta, or percreta (Fig. 4B); and showed a normal cervical length. The proximal two thirds of the vagina had thickened walls from prior surgical repair, and the distal vagina

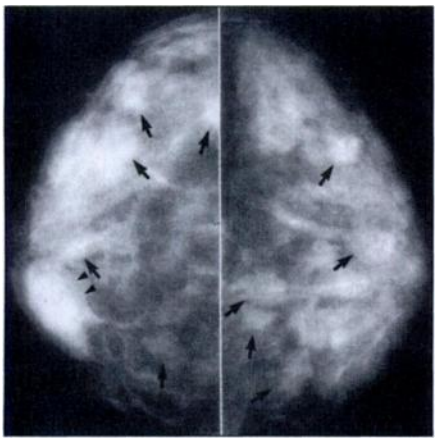

Fig. 5.—Craniocaudal mammography of 42-year-old woman shows multiple, noncalcified, poorly defined soft-tissue nodules (*arrows*) in both breasts. Largest one is seen in right subareolar area (*arrowheads*).

was atretic. MR imaging confirmed the shared opening of the urethra and vagina (Fig. 4C). Abdominal MR imaging revealed a normal left kidney and an atrophic right kidney filled with fluid, lacking surrounding renal parenchyma, and communicating with a dilated right ureter (Fig. 4D). The bladder was elongated.

After an uncomplicated pregnancy, at 38 weeks' gestation an elective low-transverse cesarean delivery was performed. A healthy boy weighing 2.6 kg was born, and the hospital course was uneventful.

This patient with McKusick-Kaufman syndrome had postaxial polydactyly and hydrocolpos associated with an inadequate urethral opening and a vaginourethral communication, a usual finding for this syndrome [3, 4]. MR imaging not only helped better delineate the patient's aberrant anatomy but also helped with the selection of an appropriate treatment plan—namely, to allow the patient to continue to carry the fetus to near term without precipitous intervention for abnormal placentation. MR imaging also showed the likely pathway necessary for successful fertilization: a common vaginourethral opening, a narrowed distal third of vagina and a fully patent proximal two thirds, and a normal cervix.

This case is important not only because the patient survived but also because she had a nearly full-term gestation and subsequently bore a healthy baby—probably the first case of pregnancy after corrective surgery for McKusick-Kaufman syndrome.

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Multicentric Granulocytic Sarcoma of the Breast: Mammographic and Sonographic Findings

Granulocytic sarcoma (chloroma) has been recognized for many years as a rare manifestation of acute myeloblastic leukemia [1–3]. Only a few cases of mammary granulocytic sarcoma have been reported, either antedating or coincident with acute leukemia [1, 2, 4]. Granulocytic sarcomas can occur at any age and at various sites in the body and frequently are multiple [1, 2, 4]. We present a case of multicentric granulocytic sarcomas in which mammography and sonography were performed and a diagnosis was made by operation.

A 42-year-old woman presented with a 2-week history of mass palpation in the subareolar area of the right breast. Physical examination revealed multiple palpable nodules in areas of both breasts in addition to the right subareolar area. Enlarged axillary lymph nodes, skin changes, and nipple retraction were not found. Both mediolateral oblique and craniocaudal views showed multiple circumscribed or indistinct nodules in both breasts, with the largest nodule in the right subareolar area (Fig. 5). Sonography revealed variably sized multiple homogeneously hypoechoic nodules, hypoechoic nodules with central echogenicity, and a heterogeneously hypoechoic mass (Figs. 6A–6C). Several nodules were excised. Histopathologic examination showed numerous immature blasts, and the results of cytochemistry were consistent with granulocytic sarcoma. Bone marrow biopsy showed more than 90% cellularity with diffuse infiltrated blasts and confirmed acute myeloblastic leukemia.

Hematologic malignant tumors can occur in the breast most often with lymphoma, less frequently with leukemia, and rarely with myeloma. Lymphoma or leukemia may infrequently be a primary breast tumor but usually involves the breast as part of a diffuse, multicentric, or disseminated process [1, 2].

Chloroma was first described by King [5] in 1853 and was so named because of its color. Myeloperoxidase in the tumor cells accounts for the green color of the tumor. But the term "granulocytic sarcoma" is currently preferred. Granulocytic sarcomas are reported in 3–9% of patients with myeloblastic leukemia and occur at the same time as, after, or (rarely) before the onset of leukemia [2, 4]. Barloon et al. [2] noted multiple, noncalcified, irregular masses

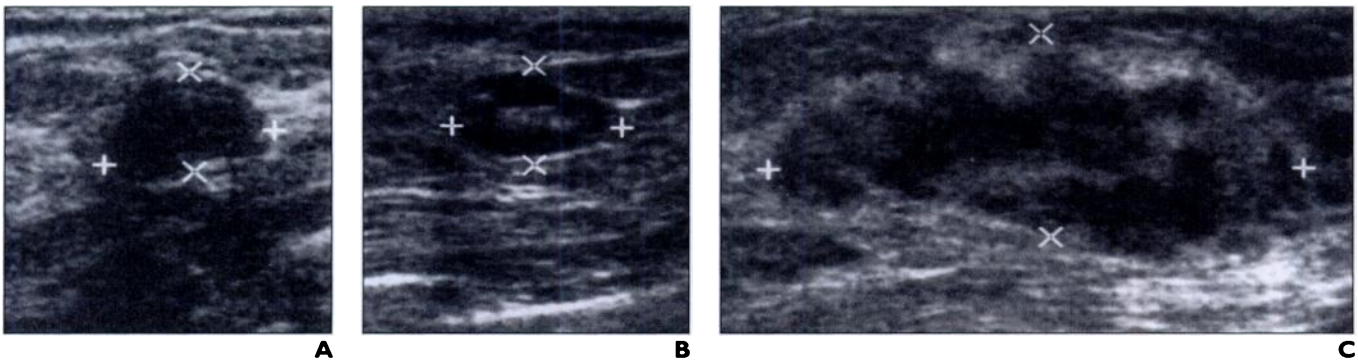

Fig. 6.—Breast sonography of 42-year-old woman.
A, Small, homogeneously hypoechoic round nodule is seen in right outer central breast.
B, Small, hypoechoic round nodule with central hyperechogenicity is seen in left upper outer breast.
C, Macrolobulated, 3.5-cm, heterogeneously hypoechoic ovoid mass is seen in right subareolar area.