ABSTRACT
Vaginal agenesis seen in Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a condition with significant psychological and social consequences where there is embryonic underdevelopment of müllerian duct. It is estimated to occur one in 4000 to 5000 live births. Normalizing sexual life with vaginal reconstruction to adequate length is usually the treatment of choice. Among many non-surgical and surgical options available, McIndoe vaginoplasty is the commonest surgical management performed worldwide with various modifications, like the use of amnion as a graft. A 19-year-old girl with normal secondary sexual characters came with vaginal agenesis with hypoplastic uterus. MRI revealed streak like structure over the pelvis with imperforate hymen and atresia of the vagina. Surgical intervention included creation of vaginal canal from a potential space between urethra and bladder above and rectum below and a fresh amnion mounted mould was placed within the canal. The amnion graft was seen well taken with resultant 4 to 5 cm of vaginal canal when mould was removed 10 days later. She was counselled and taught to remove and insert the mould to prevent vaginal narrowing and maintain the length. Amnion graft vaginoplasty has with good success rate with minimal postoperative pain, infection and scarring and thus is a rewarding procedure especially in low resource countries.

INTRODUCTION
Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is the congenital condition which refers to müllerian agenesis caused by embryologic underdevelopment of the müllerian duct, with resultant agenesis or atresia of the vagina, uterus, or both. It is estimated to occur one in 4000 to 5000 live births. This poses great deal of psychological and social problem like primary amenorrhea, painful or difficult intercourse and infertility. Reconstruction of vagina to an adequate length is the mainstay of the treatment so that the patient can have a normal sexual life. Many methods for vaginal reconstruction with grafts have been described with each having its own complications and limitations. Modified McIndoe procedure using amnion graft is simple and rewarding procedure especially in developing countries.

CASE REPORT
A 19-year-old girl from Nuwakot, a district from central Nepal came to outpatient department of Kathmandu Model Hospital with complaints of inability to attain menarche and lower abdominal pain since one year. The abdominal pain was dull aching, non-radiating occurring on and off. The pain lasted for 2 to 3 days which used to get relieved with analgesics which she used to buy from nearby clinics. Her bowel and bladder habits were normal. On examination her general condition was fair with stable vitals. Systemic examination was normal. On evaluation she was found to have a normal female phenotype with well-developed secondary sexual characteristics. Her breast development was normal with Tanner staging of 4. Axillary hair was normal. Pelvic examination revealed normal distribution of pubic hair. Labia majora and minora well developed. Vaginal examination revealed imperforate hymen with a dimple over the introitus below the urethral opening (Figure 1). Ultrasonography revealed uterus like structure measuring 3.9 x1.4 x1.9 cm. Cervix and vaginal canal could not be delineated. MRI revealed streak like structure over the pelvis with imperforate hymen and atresia of the vagina. Her renal and skeletal examination revealed normal findings. All other routine investigations were within normal limits. With the diagnosis of MRKH syndrome she was planned for vaginoplasty with amnion graft.

On the day of surgery amnion was separated from a placenta after elective cesarean section. The amnion donor was screened negative for HIV, HBsAg and HCV and consent taken from the donor. The separated amnion was washed thoroughly with sterile normal saline and kept in the saline till further use.
The patient was kept in a lithotomy position. After painting and draping, Foley's catheterization was done. In the meantime, a 7 cm mould was made from foam rolled over a small plastic rod from POP plaster which was then taped and covered with two sterile condoms inserted (Figure 2).

A transverse incision was made over hymen and a potential avascular space of 5 to 6 cm was created in between bladder and urethra above and rectum below with gentle digital and dilator manipulation (Figure 3). The amnion was mounted over the mould and its ends sutured so that amnion covered all parts of the mould. The amnion mounted mould was then inserted into the newly created vaginal canal gently so that amnion don’t get damaged. The outer end of the mould was then sutured with the labia minora to hold the mould in place and to prevent expulsion (Figure 4). Post operatively patient was kept on antibiotics. Mould and the Foley’s catheter was removed on day 10 of surgery. There was 4 to 5 cm of vaginal canal created with no blood clots or signs of infection. The amnion lining had been taken up and the cavity was gently cleaned. A newly made well lubricated mould was then inserted and the patient was discharged with the advice to follow up every fortnight. Patient was taught to remove and reinsert the mould at the time of bladder and bowel evacuation. Patient was counseled about the importance and method of placement, removal and cleaning of the mould.

**DISCUSSION**

The commonest cause of vaginal agenesis is MRKH syndrome where there is aplasia or severe hypoplasia of vagina resulting in a vaginal dimple with variable depth and with variable development of uterus. It may be isolated or sometimes associated with renal or skeletal abnormalities. Patients
with MRKH syndrome have a normal female karyotype with normal ovaries and ovarian function, thus they develop normal secondary sexual characteristics. Most of the patient have rudimentary uterus which are nonfunctional while 7-10% may have functional endometrium.

The principal aim of the treatment is to restore the sexual function by creating a vaginal canal at the normal anatomical position and axis. This neovaginal canal is created within an avascular space between the urethra and bladder above and the rectum below. Various non-surgical and surgical methods have been described. Non-surgical treatment includes self-dilatation with graduated dilators as described by Frank which may take months to develop a canal. Success depends upon the counselling, compliance and determination of the patients for ongoing dilatation.

The timing of surgery when performed depends upon the need of the patient and the type of procedure planned. Surgical procedures are often performed in late adolescence or young adulthood when the patient is mature enough to agree to the procedure and to be able to adhere to postoperative dilatation. McIndoe operation has been the commonest and the successful surgical procedure for creating neo vagina yielding satisfactory results as shown by Klingele et al improving quality of life and sexual satisfaction and provides a functional vagina with minimal complications. Original McIndoe procedure describes the use of split thickness skin graft in the newly created vagina but several investigators have sought for the modification on the use of graft like peritoneum, buccal mucosa, intestines, amnions etc.

Amniotic membrane has been used in clinical medicine for a long time starting from the use as a skin substitute to uses in ophthalmic surgery, and now with uses in wound care, orthopedic, and various other disciplines like gynecology. It is very cheap and easily available and its anti-inflammatory and antiscarring properties helps in regenerative and healing processes. Acute immune rejection does not occur after the transplantation of human amniotic epithelial cells because they do not express any antigenicity like HLA-A, B, C, and DR antigens on their surfaces.

After Brindeau used amnion graft in Mullerian agenesis in 1934, its use has been widely popularized and reported good success by many investigators. Morton and Dewhurst reported more than 90% success rate when they operated with amnion graft in 27 patients suffering from MRKH syndrome. Similarly Sharma et al after performing amnion graft vaginoplasty in 17 patients showed 88% success rate. Both the investigators reported complete epithelialization and metaplasia of the amnion into squamous cells. This provides a natural layer for wound healing and contains various important growth factors and biological macromolecules important in wound healing so that postoperative scarring, pain and infections are also less than that seen in skin graft due to its anti-inflammatory properties.

After 2 weeks of the surgery, our patient had 4 to 5 cm of vaginal length but the success depends upon compliance of the patient on regular self-dilatation of the newly created vagina and regular sexual activity to maintain her vaginal length and the width. The best definition of success is a vagina that is functional for comfortable sexual activity, as reported by the patient. There is no starting length associated with functional success. The psychologic effect of the condition should not be underestimated; hence the requirement of proper counselling session is essential. Apart from all these the best predictor of outcome after surgery is a good relationship between the patient and her parents or guardians and the ability to share their feelings with family and friends.

CONCLUSION

In patients with vaginal agenesis in low resource settings, a modification of the McIndoe method where amnion graft is placed over the vaginal mould can be done successfully with little chance of vaginal restenosis in the future, given the good patient compliance.

REFERENCES: