newsletter

p.a.g.e

paediatric adolescent gynaecology in evolution

2020 | Issue 1

Why do I want to subspecialize in

PAG?

Introduction to

AOSPAG

MRKH syndrome

Patient perspective and setting up support group

A publication by

ASIA OCEANIA SOCIETY OF PEDIATRIC & ADOLESCENT GYNECOLOGY
AOSPAG

IN THIS ISSUE

EDITOR'S MESSAGE

By Associate Prof Dr. Ani Amelia Dato' Zainuddin

I am greatly honoured to be given this task by the board members as Chief Editor for the AOSPAG newsletter, PAGE, which is the abbreviation for "Paediatric and Adolescent Gynaecology in Evolution". My aim is for this newsletter to bring together all those interested in female paediatric and adolescent health, from the different countries in Asia Oceania, using this platform to exchange ideas, collaborate, network, share our knowledge and experiences and uplift each other. AOSPAG hopes to establish the PAG fraternity in Asia Oceania in order for this important subspecialty in Obstetrics & Gynaecology to be recognized so that we can be empowered to promote for and advocate for the good health of our young girls.

We plan to have a theme for every newsletter with articles centered around this theme. For this first issue, we decided to make our theme; MRKH syndrome. Thus we have included an article on understanding this syndrome, the perspective of an individual living with MRKH, challenges of setting up the support group as well as the treatment modalities and a case report of an unusual finding associated with this syndrome.

This newsletter will be distributed to all members of AOSPAG every 6 months by email, there will be no cost attached nor will there be any hardcopies. Anyone wishing to contribute an article on anything pertaining to female paediatric and adolescent gynaecology are welcome to email to myself, Ani Amelia Zainuddin, at aospagpage@gmail.com. We welcome ideas and feedback so this newsletter grows and improves with each subsequent issue.

Thank you and enjoy reading!

Asia Oceanic Society of Paediatric and Adolescent Gynaecology, or known as AOSPAG, was first established in June 2017 in Hong Kong.

This is a Society established for countries in Asia Oceania to promote the subspecialty of Paediatric and Adolescent Gynaecology. The countries include the Philippines, China, Hong Kong SAR, India, Japan, Laos, Malaysia, South Korea, Australia, New Zealand, Indonesia, Pakistan, Thailand and Vietnam. We hope many more will join.

The Board Members of AOSPAG

Ma. Socorro Bernardino
President

Associate Prof Dr. Ani Amelia Dato' Zainuddin
Vice President

Dr. Symphorosa S.C. Chan
Secretary

Hideya Sakakibara
Treasurer

Sonia Grover
Education coordinator

Photo of our first board meeting on 12th June, 2020. Prof Hideya was not able to join as he was very busy with Covid-related duties in his hospital.

The board members are busy O&G consultants specializing in PAG in their respective countries. They have come together to establish AOSPAG. They believe in the importance of this subspecialty, in providing optimal health care for our girls and young ladies in all our countries to enable them to prosper and attain their best future, within our unique cultures, rich traditions, diverse religions and amazing peoples.

MISSION

AOSPAG is dedicated to be at the forefront of young girls and adolescents' reproductive health needs.

VISION

To create and foster camaraderie and teamwork among the Asia Oceanic Nations in the advocacy of PAG

VALUE

- Research Innovation
- Integrity in Training
- Dynamism in Learning
- To take Action on Environmental Issues

What we have done so far

Monthly AOSPAG webinar sessions hosted by different countries every first or second Thursday of the month, started from July 2020 by Australia, followed by the the Philippines in August, the Malaysian team in September, then Hong Kong in October, China in November and Pakistan in January 2021.

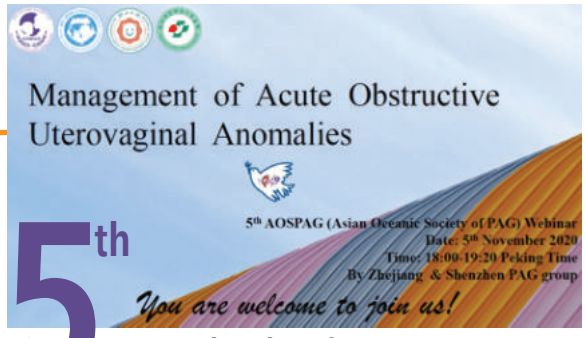
Topics already presented in the AOSPAG webinar sessions;

2nd July by Australia
Labial swelling in young girls

3rd September by Malaysia
Management of menstrual issues in adolescents with special needs

6th August by the Philippines
Growing teratoma syndrom

8th October by Hong Kong
Adolescent PCOS: Controversies and Updates



5th November by China
Management of acute obstructive uterovaginal anomalies

Newsletter PAGE first issue!

A brief history of AOSPAG

Dr. Symphorosa S.C. Chan commenced the Pediatric and Adolescent Gynecology (PAG) service in Prince of Wales Hospital, The Chinese University of Hong Kong since 2002. Her center was accreditated by FIGIJ as a training center in PAG in 2009. She served FIGIJ as a Board member from 2010-2019, was the Chairperson of the 17th FIGIJ World Congress of PAG in 2013 & served as the Director of the IFEPAG examination since 2013.

Asia Oceania Society of Pediatric and Adolesecnt Gynecology was established in June 2017.

Back to 2016, during the 18th World Congress in Pediatric and Adolescent Gynecology in Florence, Italy, Dr. BM Kang of South Korea introduced the idea of establishing the Asia Oceania Society of Paediatric and Adolescent Gynecology to participants from Hong Kong, Korea, Malaysia and Philippines. In view of the large number of Asia countries and the unique culture of different countries and difference in health care practices between Asia and the Europe, South America and North America, the whole group was eager to form our own society. It was decided that Asian Oceania PAG specialists could meet in the subsequent year during the 25th Asian & Oceanic Congress of Obstetrics and Gynaecology (AOCOG).

A group of 17 Paediatrics and Adolescent Gynaecologists coming from 9 regions of Asia, including China, Hong Kong SAR, India, Japan, Laos, Malaysia, South Korea and Thailand, gathered together on the 17th June, 2017 in Hong Kong during the 25th AOCOG. Prof. Sonia Grover of Australia also joined the meeting over phone. All participants fully supported the needs of setting up our own society with the shared objectives of gaining the recognition of this new subspecialty in Asian Oceania. She was named as Asian Oceanic Society of Paediatric and Adolescent Gynaecology (AOSPAG). The first Board was formed which included Symphorosa Chan, Ani Amelia Zainuddin, Angela Aguilar and Hafizur Rahman. Meetings in future regional events; AOCOG in Manila 2019 and WCPAG in Melbourne in 2019 were planned. More countries and members were welcomed.

AOSPAG meeting 2017

Front row (from left to right):

Liying Sun (China), Ani Amelia Zainuddin (Malaysia), Symphorosa Chan (Hong Kong SAR.), BM Kang (Korea), Apurba Bhattacharya (India), Angela Aguilar (Philippines)

Back row (from left to right):

Charleen Cheung (HKSAR), Alice Yiu (HKSAR), Takeki Tsutsui (Japan), Huihui Gao (China), Joo Hyun Park (South Korea), Nur Azurah Abdul Ghani (Malaysia), Soyun Park (South Korea), Anan Sacdpraseuth (Laos), Hafizur Rahman (India), Areepan Sophonsritsuk (Thailand)

AOSPAG meeting 2019

Front row (from left to right):

Symphorosa, Shital, Sonia, Hideya, Sox, Amelia, Ana Vetriana, Liyin Sun (Lynn), Charleen

2nd row (from left to right):

Sa Ra Lee, Takashi Takeda, Qiuxiang Shen (Fiona), Soyun Park, Nur Azurah, Gang Peng, Huihui Gao (Amy), Angela

3rd row (from left to right):

Chan, Angela, Tamara, Rebecca, Sarah

List of Current PAG Training Centers in Asia Oceania

Philippine Children's Medical Center, Section of Pediatric and Adolescent Gynecology, Philippines
Chairman: Dr. Socorro C. Bernardino
Advisory Director: Dr. Corazon Yabes - Almirante
Email: pagsphil@yahoo.com

Dept. Paediatric Adolescent Gynaecology
Royal Children's Hospital, Melbourne, Australia
Director: Professor Sonia Grover
Email: sonia.grover@rch.org.au

Department of Obstetrics and Gynaecology, Prince of Wales Hospital
The Chinese University of Hong Kong, Hong Kong SAR
Director: Dr. Symphorosa S. C. Chan
Email: symphorosa@cuhk.edu.hk

Paediatric and Adolescent Gynecology Training Center in Zhejiang University, Hangzhou, Zheijiang, China
Director: Dr. Liying Sun
Email: PAG@zju.edu.cn

Pediatric and Adolescnet Gynecology Center, Department of Obstetrics and Gynecology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea
Director: Prof. Byung Moon Kang
Email: bmkang@amc.seoul.kr

Paediatric & Adolescent Gynaecology (PAG) Unit, Department of Obstetrcis and Gynaecology, The National University of Malaysia (UKM) Medical Centre, Kuala Lumpur, Malaysia
Head of Unit: Associate Prof. Dr. Ani Amelia Zainuddin
Head of Department: Prof. Nur Azurah Abdul Ghani
Email: amelia@ppukm.ukm.edu.my; aniameliaz71@gmail.com; nurazurahag@gmail.com

Professor Sonia Grover is the Director of the Gynecology Division at the Royal Childrens Hospital Melbourne , Melbourne, Australia, Professor in Obstetrics & Gynaecology at University of Melbourne , Distinguished Professor for the National University of Malaysia (UKM) and the immediate Past President of FIGIJ (International Federation of Infantile and Juvenile Gynecology).

Introducing and Continuing Education on PAG

My enthusiasm and determination to make sure that we teach about PAG means I am determined to take every opportunity that is available. It seems an age ago that we used to be able to travel and visit, stay for a few days, teach, be involved in challenging clinical cases and do more teaching. Even the luxury of a conference/ congress seems a privilege and it certainly was a privilege to be involved. Now, we need to turn to other ways of sharing and teaching. In running the world congress here in Melbourne, we had tried to ensure that we provided all sorts of opportunities; from young women presenting, to debates, and thought-provoking lectures.

But now we need to turn to other options, so webinars, and online tutorials are all happening.

But more than that, we have the additional opportunity of working together within this organisation, with our friends and colleagues in our region. We can think about not just teaching but also trying to better understand problems, to share research opportunities.

There are a number of problems that differ in different settings. From a simple problem like heavy periods;

1. Why do adolescents present with this in some places and not in others?

2. Are there different rates of bleeding disorders in different ethnic groups?

3. By not intervening and treating heavy periods, do young women end up with other problems?

To a problem like period pain;

1. Why do so many adolescent girls attend clinics with period pain in Australia, and this doesn't happen in other places?

2. Does this mean we see less severe endometriosis because it is diagnosed earlier?

And questions like using dilators for vaginal agenesis - is that feasible in all cultural groups? I had always thought that this was not really an option in many Asian countries, yet my colleagues in Hong Kong have shown that my presumption was wrong.

What questions could we tackle and try to answer by teaming up, by contrasting and comparing experiences in different countries in our region?

I would love to hear your thoughts. And I would love to work with you.

Weekly webinars in the form of clinical case discussions, mostly relating to cases we have seen during the recent weeks, are occurring most weeks (If you are interested in joining these – please email me!). And there are the once monthly ASOPAG webinars. So far we have had a few; the first from Australia – on vulval swellings, and the second was coordinated by PAGSPHIL – on growing teratoma syndrome – I learnt so much.

Please join us!

We'd love to join our happy, friendly crowd of clinicians with an interest in PAG.

PEDIATRIC AND ADOLESCENT GYNECOLOGY:
A Perspective

Ma. Socorro Bernardino, M.D.
President, AOSPAG

There are many subspecialties in the field of Obstetrics and Gynecology, most of which deal with the different facets of reproductive concerns in adult women. The young patient is entirely different in all aspects from that of the adult gynecologic patient. Through the years, gynecologic conditions in children as well as in adolescents has emerged in peculiar manifestations that required more knowledge and understanding of the age group. Hence, the commencement of a specialized field to address gynecologic needs of this cohort - that is Pediatric and Adolescent Gynecology.

The clinical mindset and approach, the more extensive evaluation, the consideration of the psychosocial aspects, the maximization of conservative management are principles seen in the field of pediatric and adolescent gynecology. Deeper understanding of the physiology and anatomy and growth and development at par with age is necessary to formulate appropriate diagnosis and treatment plans. Some cases would require a multidisciplinary approach. Dealing with kids and teens necessitate more time and effort in obtaining a history which in children comes from parents/caretakers and in adolescents an additional consideration of confidentiality. Awareness of the legal statutes, family dynamics, and cultural boundaries is also of importance. The gynecologic evaluation and examination in children is also a noteworthy consideration. Menstrual aberrance and concerns differ in cause and treatment. Trending issues on teen pregnancy, STI, and contraception are also on the rise that need to be directed. Surgical management requires a thorough reflection on the appearance of pubertal development in children and reproductive health and future fertility in teens. These are aspects that make PAG unique and dynamic.

The realization of Pediatric and Adolescent Gynecology in the Asia-ocenia region is but young. More work is needed on baseline statistics, sharing of cases, networking in training, promotion of research and continuing medical education. As of present time, we have had 5 monthly webinars and will still continue, on the development of our statutes and have reinforced plans in participating in international symposia and with FIGIJ. On behalf of the present AOSPAG board, with ONE objective in mind – our girl patient and her future- we enjoin the whole asia-oceania community to participate and collaborate with us.

Why would you want to specialize in PAG?

Associate Prof Dr. Ani Amelia Dato' Zainuddin is the Head of the PAG Unit in HCTM UKM. She and her colleague, Prof. Nur Azurah Abdul Ghani, established the first PAG unit in Malaysia in 2008. Her special interest is in managing patients with Disorders / Differences of Sex Development (DSD) such as Rokitansky (MRKH) syndrome, XY females, patients with Congenital Adrenal Hyperplasia (CAH) and patients with Premature Ovarian Insufficiency (POI).

Why did I choose to subspecialize in PAG?

My passion for Paediatric and Adolescent Gynaecology (PAG) started when I was studying for my Masters in O&G final clinical examinations. I loved reading up about Turner's syndrome. I was fascinated by what were the other causes of primary amenorrhoea. I knew then that this was what I wanted to do. I liked young children and liked a little bit of surgery and loved labour room. So being able to be given the opportunity to do PAG and still deliver patients as an O&G specialist is wonderful.

Prof Nur Azurah and myself were the first ones in Malaysia to be trained in this subspecialty and we established the first PAG unit in April 2008. Prof Nur Azurah went first to Melbourne to train under Prof Sonia then I followed, doing my PhD in outcomes of female patients with Congenital Adrenal Hyperplasia (CAH), these patients are categorized as Differences of Sex Differentiation (DSD). I found I was fascinated with patients with DSD conditions.

When Azurah was away training in the early years, I ran the PAG clinic by myself and found myself frantically looking through the short notes textbook on PAG whilst the patients were preparing themselves to be examined by me behind the curtains. I made friends with the paediatric endocrinologists, the paediatric surgeons, the general paediatricians and the psychiatrists, who were all wonderfully helpful and became my network and an integral part of the multidisciplinary team for managing PAG patients. I harassed one of my radiologist colleague to interpret MRIs of Mullerian anomalies for me so frequently that she became interested in it herself and went on to specialize in it.

I was fortunate that I was trained by Prof Sonia Grover who is a regional expert in PAG. She was also my supervisor for my PHD and is a truly wonderful mentor. I was blessed that my Heads of Departments always were supportive of the PAG unit and its development. I am blessed too to have Azurah as my partner in the PAG Unit, it is easier for two people to develop a new unit rather than a single person, however, of course, you need to get on fairly well with your partner, which we do, thankfully.

When doing PAG, I realized how important this subspecialty is. That the patients and their families often suffer from delayed or wrong diagnoses, suboptimal care and are frequently neglected due to lack of awareness of the rare conditions, not only by the public but also by the healthcare providers themselves. PAG is given scarce attention in the syllabus of many medical schools and is largely an unrecognized subspecialty within the fields of Obstetrics & Gynaecology and Paediatrics. It is thus important that I do my part in increasing the awareness of this subspecialty within the national and international fraternities.

Our unit has been accredited by FIGIJ to be an international training center for PAG, so we train both national and international fellows. I am excited to be part of AOSPAG, to work together with Dr Socorro Bernardino, an equally passionate advocate for young women, and with our committee to further expand PAG awareness and training in the Asia Oceanic countries. I plan to continue working hard and advocating for the good health of our girls and young women, God Willing.

Dr. Sachedina is born and raised in Vancouver, Canada where she also completed medical school and training in Obstetrics & Gynaecology at the University of British Columbia. She recently completed fellowship training in Paediatric & Adolescent Gynaecology at the Royal Children's Hospital in Melbourne, Australia and is soon to begin her career as a consultant Obstetrician, Gynaecologist and Paediatric Gynaecologist back in Vancouver.

The Pursuit of a Career in PAG

For too long young women have been an underserved population in medicine. Gynaecologic considerations in this group are poorly taught in medical school and in postgraduate training programs. They are normalized and dismissed by society; often considered an occupational hazard of being female. It was a desire to be a part of the movement to address this underserved population that drew me and continues to draw me to the discipline of Paediatric & Adolescent Gynaecology (PAG).

I have had the privilege of mentorship from some of the global pioneers in PAG. Their dedication, passion and achievements make me excited to be a part of this young field, which is ever growing and changing. I hope to continue, as they have, to push boundaries and re-define gynaecologic care for young people.

On a professional level, PAG is a unique field for many reasons, but perhaps most of all because of the diversity of clinical and social situations that we encounter and uncover, offering an opportunity for lifelong learning and growth. To be an effective PAG clinician, we must have the patience and attention to detail of a paediatrician, the compassion and resourcefulness of a social worker, the ability to listen of counsellor, the precision of a surgeon, and the wisdom to know when to ask for help.

Challenges in setting up Paediatric and Adolescent Gynaecology services in Pakistan

Dr. Iffat Ahmed is Assistant Professor in Obstetrics & Gynaecology at Aga Khan University, Karachi, Pakistan. She completed her fellowship in PAG at UKM and is now busy setting up services in PAG in her university, She is the pioneer of PAG in Pakistan.

The history of the development of Paediatric and Adolescent Gynaecology (PAG) services in Pakistan is very short. My PAG fellowship training, from University of Kebangsaan Malaysia (The National University of Malaysia), completed earlier this year 2020 in Feb, and I returned with an intention to commence not only sub-specialty services at The Aga Khan University (AKU), but also to establish the first formal training of PAG in Pakistan.

Apart from many expected challenges such as staffing, logistical and financial considerations, due to the unforeseen COVID-19 pandemic outbreak, the hospital had to limit out-patient services. Overall patient load were reduced to urgent cases only and only a limited number of clinics were allowed to run, therefore the designated PAG clinic could not be started. The rigorous process for credentialing at AKU was further delayed by the pandemic. Although the university and department of ObGyn provided logistic support for provision of clinical space and operation theater slot, but the census is still not at par as it used to be.

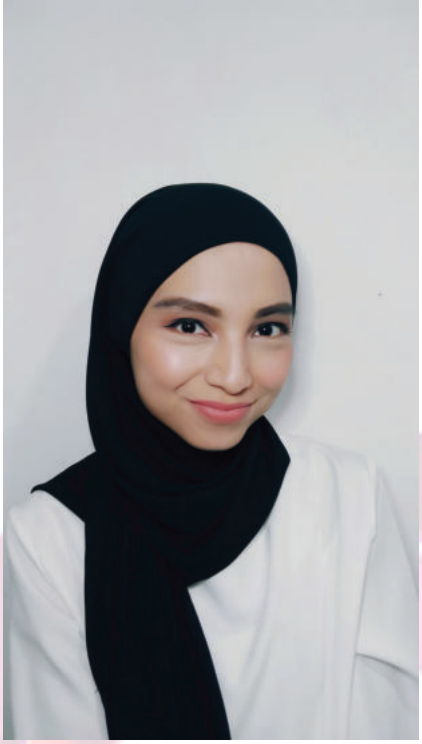
PAG is not a well-known sub-specialty in Pakistan, moreover our referral system is not yet well established. As an initial strategy for developing awareness among physicians, I organized webinars on introduction to PAG and management of sexual assault among children and adolescents. I am grateful to colleagues from Malaysia for their unconditional support by their participation in the webinars.

Further on the educational front, I developed a module which was executed for post-graduate ObGyn trainees. This module was designed on student-centered spiral-curriculum which included student presentations, workshop on management of amenorrhea and guest-speaker lectures.

The female child, within the social structure found in Pakistan, especially from the marginalized groups, receives the minimal share of the family income, for her food, education and health. Seeking medical help is thus far less likely to occur when it comes to her reproductive health. Public awareness sessions which can reach out to the communities are the way forward, along with strategic collaborations and mutual referrals within the fraternities of Paediatric surgery/endocrinology, adolescent medicine and psychiatry.

My PAG services are based in Aga Khan University (AKU), which is a private organization. An average Pakistani family will not be able to bear the high medical expenses if they seek PAG services despite having some financial support. Therefore, the ultimate challenge would be to provide PAG services to grass root level by involving and mobilizing the government sectors. Thus, I need to increase awareness of PAG in Pakistan and enable the training of future specialists in PAG who can serve the government hospitals and clinics. I have already taken the first few steps in doing so and will strive on.

LIVING WITH MRKH



Wani Ardy is a Malaysian published author, poet-performer, singer-songwriter, and an active MRKH advocate.

She founded MRKH Malaysia in 2014; a support group empowering Malaysian women with MRKH Syndrome, as well as their families.

Wani has been creating awareness through numerous media platforms and she represented Malaysia at the Global MRKH Conference 2019 in Australia by presenting the works that she has done for Malaysian MRKH community.

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At the age of 36, I did not think there will be anything new that MRKH could possibly bring to my plate. I thought that was it; I was done, I have passed all the bumps in my life as an MRKH woman. Diagnosed at 17 years old was a pretty big hit for me. I was young and raised in a conventional Asian Muslim family. All I wanted to do was to fit in like other normal teenagers. I did not want to be different. I did not want to feel left out or isolated by not having periods and not having friends to relate to. I definitely did not want to be 1 in every 4,500. Unlike my peers who were worrying about homework, final exams, and life after school, I spent the last bit of my adolescence worrying about my fast forward future – acceptance or rejection, marriage or spinsterhood, monogamy or polygamy, babies or childlessness – when I myself was just a kid. But the real challenge was enduring my 20s. Nearly every year I would get admitted for abdominal pain and UTI. I discovered the term MRKH Syndrome and dilation therapy upon googling.

I also discovered that not many men are alright with their partner not being able to carry children. One after another failed relationship heightened my battle with self-worth and led me to major depression. I was married for almost a decade and came out of it divorced, with PTSD and psychosis. Despite all these, I have no regrets. I would not be my son's mother and the advocate I am today if it was not for these experiences. This year I found out that I have lost 40-50% of my hearing. It impacts my work as a poet performer and a singer-songwriter, and I have been struggling. But slowly I am learning to read lips, facial expressions, hand gestures, body language, and work with my muscle memory. I have also accepted that families and friends - or even the society - could never truly understand, and that's okay, so long as I have the support of my MRKH community. As always, I shall try my best to soldier on because I know this journey is full of tests. I choose to be grateful for everything that I do have. MRKH is a lifelong gift. It does not in any way define me. It strengthens me. God-willing.

THE CHALLENGES IN SETTING UP THE MRKH MALAYSIA SUPPORT GROUP

By Wani Ardy

It was a naïve dream. I didn't want other MRKH girls to feel as lost and lonely as I was growing up. That was why I mustered up the courage to begin talking about my condition publicly, through my writings and performances.

It was not easy. My parents were conservative. My father was an imam and my mother explicitly told me not to tell anyone that I could not carry any children. Even when I adopted my son, my mother would advise me to not tell my friends the truth, to tell them that this baby was from my (nonexistent?) womb. Of course, it was impossible. I went behind my parents' back anyway, but I kept praying to God to guide me throughout my journey as an advocate.

I was lucky that I've built up my name as a published author and performing artist at that time, so I was not starting from scratch in terms of reaching an audience. But it also meant that now the whole country knew that I, Wani Ardy, am the girl without a womb. It's a heavy 'label' and sometimes it feels like I have that label across my forehead.

It was during one of my shows when my first ever Malaysian MRKH sister came and approached me. And it was overwhelming. Just the feeling of being able to talk face-to-face, look and hold this person who was in my situation– we instantly cried upon that first meeting. From that moment on, I decided to search for more MRKH girls and women. I created a support group on Facebook in 2014 upon resigning from my full-time lecturing job and I named it MRKH Malaysia. From just 2 members, we slowly grew into having almost 200 members today, which to me is a big number for a rare syndrome.

Setting up and managing MRKH Malaysia of course has its challenges. Maybe I'm not able to describe them all out here, but I would say there were five main challenges throughout my experience as an ever-learning founder:

First, verification. When I initially started the support group on facebook, it was open to all. Then I realized that we began to have people who were non-MRKH joining us. We also had women who pretended to have the syndrome, all because they were curious and intrigued- they wanted to be part of something that they thought was interesting and unique. Many MRKH women weren't happy about this as our discussions were very personal and MRKH-related, that eventually I had to change the setting of our support group to closed and private. We also request for medical report or doctor's letter that states an MRKH diagnosis as a form of verification. This sounds like a hassle to many people who applied for membership, but we need to be cautious and specific for the sake of our existing members. The only non-MRKH people that we do accept are female guardians (e.g. mothers, sisters) to MRKH girls who are 18 years old and below.

Second, confidentiality. The majority of the Malaysian MRKH women are very secretive about their condition and identities. They have a lot of shame and guilt, and it's not something that they're proud to share. I believe this is normal, sadly, for most Asian MRKH women due to our conservative culture and tradition. Many of them keep their diagnosis a secret even from family members and close friends. It is my responsibility to keep it that way in order to gain their trust and ensure their comfort of being in the support group. I do a lot of advocacy work on social media to spread awareness, so when I post photos of them online, I've learned to 'blur' their faces out of respect.

Third, participation. Because of the 'reserved' nature of Malaysian MRKH women, it's pretty hard to gather them all. Although there are almost 200 of us, every gathering consists barely 20. This makes it a bit difficult for me to hold an event specially for them. Most don't turn up and frequently change their minds at the last minute. Majority just aren't ready to meet each other eye to eye in a group and prefer not to face the issue upfront. Many of them, however, do communicate with me individually quite often. We would meet somewhere privately and sometimes their spouses or mothers come along too, and they are comfortable to ask me more questions in person.

Fourth, dealing with media and cultural stigma. Fact: Malaysian media loves sappy stories and anything controversial. I've learned this the hard way and I've learned my lesson. Solution: I hand my list of terms and condition whenever a radio station, tv channel, online or printed media wants to interview me or feature MRKH-related content on their programs. I would talk to the producers and make sure that the presentation of the content will be empowering and positive instead of sad and negative. I also prohibit them from using my personal photos or anything that involves my family or my history, because the spotlight should be on MRKH Syndrome and MRKH Malaysia, not the founder. I need to always cross-check every article and every script because I don't want our society to get the wrong idea or any false information.

Fifth, staying sane as a founder. I won't lie, I have my moments of breaking down. It's tough when your face is the only one out there. I still get anxiety attacks whenever I see my story in the newspaper or on tv. I imagine my parents and in laws reading them, watching them. It doesn't get easier. But, in the end of the day, it feels worthy because every coverage brings more Malaysian MRKH women to our support group and our community grows stronger. The most important thing is that we've found each other.

UNDERSTANDING MRKH

MRKH syndrome is a rare condition affecting 1 in 4,000 to 5,000 newborn girls. In most cases, they have normal karyotyping of 46XX with normal functioning ovaries hence have normal female secondary sex characteristics. The most common clinical presentation is primary amenorrhoea. Vaginal maybe partially present or completely absent with uterine abnormalities that ranges from a rudimentary to an absent uterus.

MRKH occurs sporadically but it has been reported in families hence suggest an autosomal dominant inheritance with reduced penetrance and variable expressivity either due to single gene or limited point mutation. The risk of recurrence is estimated to be 1-5% in first degree relatives. Several genes have been identified to cause MRKH such as LHX1, TBX6, WNT9B and WNT4.

The MRKH syndrome can be classified into two subtype: isolated/typical (Type I MRKH) or atypical/ in association with extragenital malformations (Type II MRKH). In the isolated type, there is absence of uterus and upper part of the vagina without any other extragenital malformation. The atypical type is associated with abnormalities of the kidneys, spine, ear and heart. Abnormalities of the kidneys include unilateral renal agenesis, ectopic kidney, horse-shoe kidney and hydronephrosis. The most common spine anomaly is scoliosis. Those who experience hearing problem is most likely due to conductive defects such as stapes fixation or sensorineuronal deafness. They may also have digital abnormalities such as polydactyly or syndactyly. Mullerian hypoplasia, renal agenesis, cervicothoracic somite dysplasia (MURCS) association is the severe form of type II MRKH.

Previously, diagnostic laparoscopies were performed to confirm the diagnosis. Nowadays, laparoscopy procedures are reserved for surgical interventions. Magnetic resonance imaging (MRI) has a high sensitivity and specificity when reported by radiologist who are familiar with these anomalies. MRI also offers an advantage of assessing the urinary tract and the spine for any abnormalities. Hence, MRI is now considered the gold standard for diagnosis of MRKH. Nevertheless, pelvis ultrasound is a good modality especially performed by trained personnel in low resource countries.

Prof Nur Azurah Abdul Ghani is the Head of the Department of Obstetrics and Gynaecology, UKM Medical Centre, Kuala Lumpur. Was the first to be trained in PAG in Malaysia. Together with her colleague, A/Prof Dr Ani Amelia Zainuddin, they set up the first PAG clinic in Malaysia. The centre is now recognised by FIGIJ as one of the training centre for IFEPAG fellowship.

Too often the diagnosis of MRKH is missed and patients may end up with unnecessary surgical procedures. This, no doubt would create anxiety and frustration not only to the young girl but also her parents. Clinicians should portray more empathy during the breaking of the news as this may have a devastating impact of the life of this young girl. Too often the girl is told that she has no uterus and unable to bear a child. The girl might feel depressed, confused, shocked, different, have a fear of rejection and low self-esteem upon hearing the diagnosis. Referral to a psychologist may be necessary.

In principal, the creation of a vagina should be carried out only when the girl is emotionally mature and mentally ready to embark on a sexual relationship. Many would agree that non-surgical approach should be offered prior to surgical intervention. Frank's method using vaginal dilators has been shown to have a success rate of more than 70%. There are many surgical methods of creating a neovagina which include McIndoe, Davydov's, Vecchietti, Luohu's operation and also using bowel as vagina. The advantages and disadvantages of each method need to be explained to the patients.

Recently, uterine transplantations have resulted in successful pregnancies and live birth babies. This no doubt has given hope to women with MRKH globally. Nevertheless this is this at experimental stage that requires strict selection criteria.

Girls with MRKH are often felt traumatised by the diagnosis hence clinicians have to show more empathy and be compassionate upon revealing the diagnosis.

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LUOHU VAGINOPLASTY FOR MRKH PATIENTS

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Dr. Pan's clinical expertise lies in the treatment of MRKH syndrome, congenital uterus cervical atresia, and other kinds of female reproductive tract malformations.

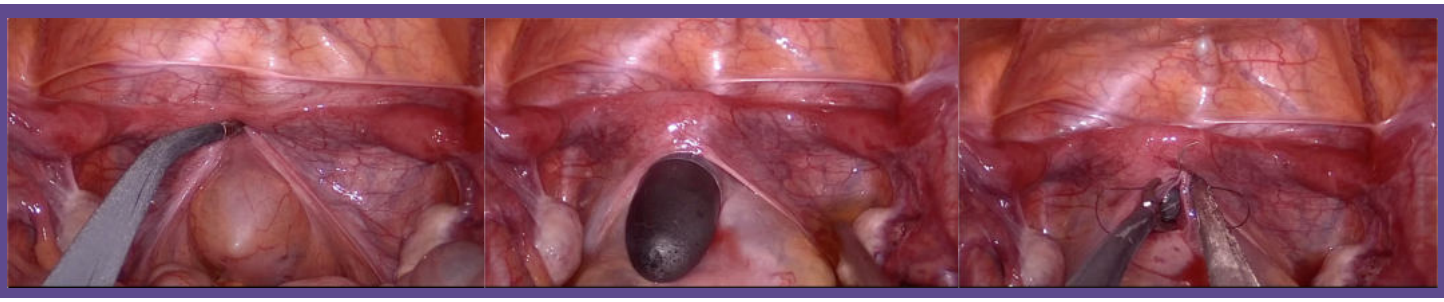
Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, or vaginal agenesis, has an incidence of 1 per 4,500–5,000 females. Patients will be able to achieve anatomic and functional success by surgical intervention. In cases in which surgical intervention is required, referrals to centers with expertise in this field should be considered, as few surgeons have extensive experience in construction of the neovagina and its relevant surgery by a trained surgeon offers the best opportunity for a successful result.

Surgical procedures usually are performed in late adolescence or early adulthood when the patient is mature enough to undergo the procedure and persist in postoperative dilation.

The Luohu vaginoplasty is the fruit of our team's experience and is developed as a three-stage operation. Our team have already performed this procedure on 1000 MRKH patients since 2001. A systemic approach to Luohu vaginoplasty for MRKH syndrome is as follows:

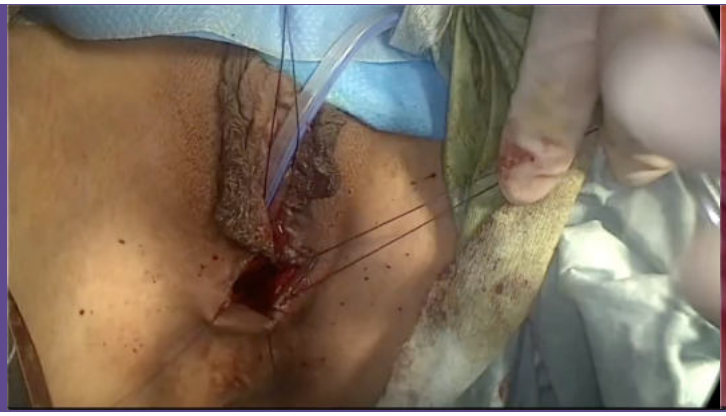
Stage 1.

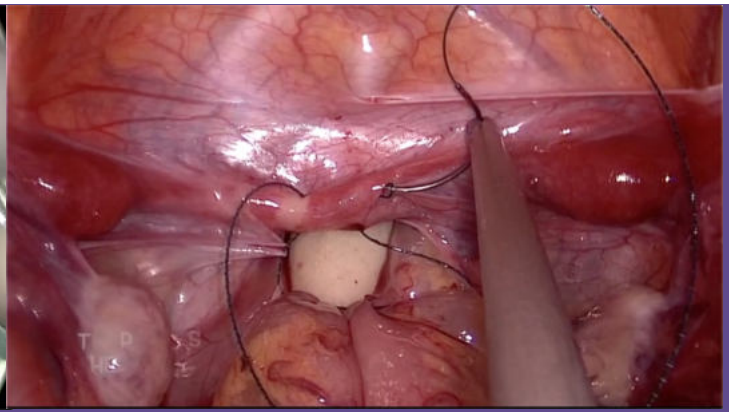
Laparoscopic part, formation of a neovaginal canal between the bladder and rectum. a Veress needle is inserted through the gap between the bladder and rectum. Hydrodistension is created by injecting normal saline with diluted adrenaline. The distended peritoneum is then isolated from the rectal wall. The needle pierces through the thin peritoneum. Per rectal examination is done to exclude rectal injury. Laparoscopic suction catheter is inserted to reach the vaginal needle, and then the needle is replaced by a medium sized curved clamp, which is pushed up along the suction catheter into the peritoneum. Opening the forceps will further enlarge the tunnel. Vaginal dilators from size no. 1 to 6 are used to dilate the neovaginal canal to a diameter of 3.5cm. Four stitches are applied to hold the peritoneum of neovaginal vault at 3, 6, 9 and 12 o'clock positions respectively. Each needle is retrieved vaginally and secured outside at the perineum.



Stage 2.

Vaginal part, fixation of the distal end of the peritoneum in place of the vestibular mucosa. The needle at 6 o'clock is sutured to the mucosal junction at the perineum. Subsequently we stitch and hold the remaining sutures to 3, 9, 12 o'clock of the perineual mucosa correspondingly. When stitches at all positions are held in place, we start tying the peritoneum and mucosa from 6 o'clock. The knot is pushed deep into the neovaginal canal. The peritoneum and the mucosa are facing each other. The perineal mucosa is brought up into the neovaginal canal due to tension.

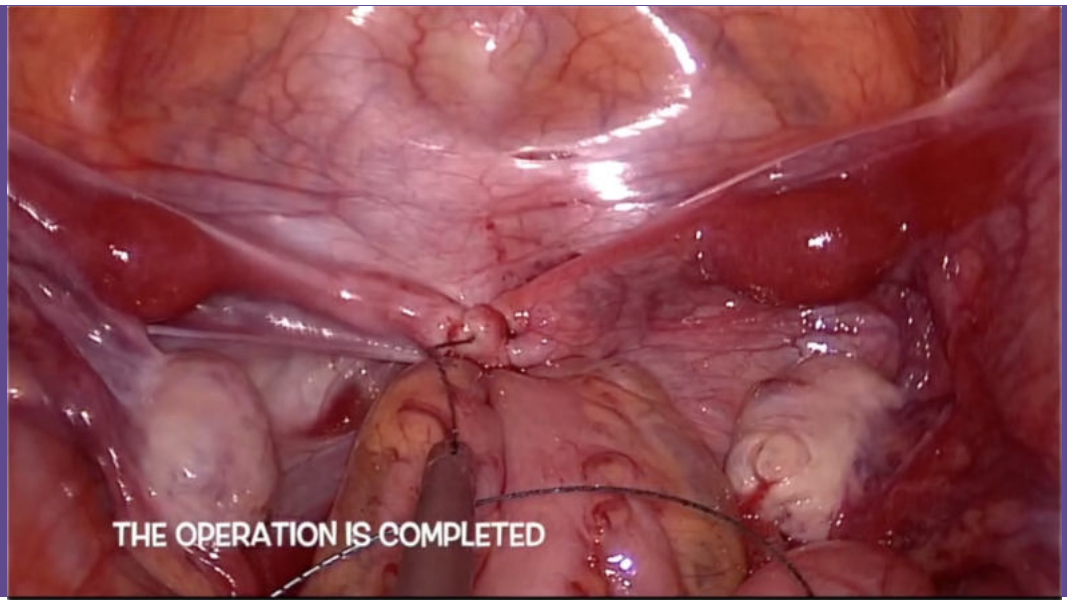




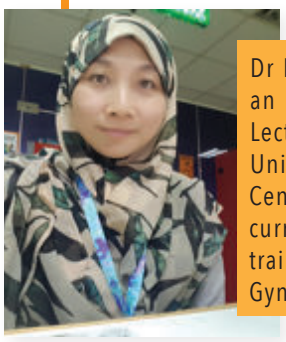
Stage 3.

Laparoscopic part, closure of the proximal peritoneum in the pelvic cavity. A barbed suture is used to close the pelvic peritoneum with a seven-stitch purse string technique.
A vaginal gauze is inserted into the neovagina and will be removed 5-6 days after operation.

Postoperative dilation is essential to prevent obvious neovaginal stenosis and contracture; instead of long-term implantation of vaginal mold, patients were treated with postoperative vaginal dilation. Vaginal dilation is recommenced to perform 5-10 minutes on a daily basis. Frequency of dilation can be reduced after patients become sexually active.



AN ALTERNATIVE APPROACH TO NEO-VAGINAL CREATION IN YOUNG WOMEN



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Dr. Anizah Ali is a Consultant in Obstetrics and Gynaecology and completed her fellowship in PAG at UKM this year. She will be embarking on her PhD journey soon , investigating patient and healthcare workers' acceptance and perceptions on fertility preservation.

Vaginal dilatation therapy is a non-invasive therapy advocated for females with pathologies presenting with inadequate or non-existent vagina. Among such pathologies are the spectrum of Mullerian anomalies i.e Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH), Vaginal agenesis or atresia, obstructed hemivagina with ipsilateral renal agenesis (OHVIRA), and transverse vaginal septums (TVS). This vaginal therapy is also used to increase the vaginal width in cases of vaginal narrowing or strictures following surgeries as well as prophylactically post-neo vaginal creation to maintain vaginal patency.

As a first-line therapy for neo-vaginal creation in cases of MRKH, vaginal dilation therapy has the advantages of being non-invasive with excellent outcomes; quoted success rates 70- 90%. Post vaginal corrective surgeries in Mullerian anomaly cases, it is an invaluable tool to ensure vaginal patency. Unfortunately, among pre-pubertal and adolescent girls this promising option may not be appealing for various reasons. Ranking highest on the list is the inadequate or misguided understanding of both the patients or parents/caretakers on the need for dilatation either as a primary therapy or as adjunct post-primary surgical repair. Secondly, within the confines of religion and culture, it may be considered taboo for pre-pubertal and adolescent girls to be introducing objects into their private part let alone dilators. Furthermore, for some Asian pre-pubertal and adolescent girls, coital vaginal dilation is not an option to be considered at this age. Lack of motivation and drive along with poor compliance hinders the success of this therapy.

Hereby, we share our centre's experience in intensive vaginal dilatation therapy among adolescents and young female adults. There were total of 5 cases; 3 were Malays, and 2 Chinese. The cases were 2 of vaginal dysgenesis, 2 cases of MRKH and one TVS.

Three were performed as adjunct following vaginal reconstructive surgery and two cases was primary therapy for neo-vagina creation in MRKH. Ages ranged from 14 to 28 years old. All of them have had prior trial of self vaginal dilation at home which failed. They were admitted for 3 to 4 days duration. Upon admission, both the dedicated intensive vaginal dilators team members briefed and counselled in length regarding the overall intensive dilatation therapy and stressed the importance of compliance to ensure successful outcome. Stepwise approach in pain management during dilation was utilised. Dilators of various sizes were used and tailored to each patients' need. Adjuvant treatment were utilised consisting of oral Paracetamol and topical application of estrogen and Lignocaine gel.

Upon commencement of therapy, the mean vaginal length was 2.12 cm with mean width of 1.65 cm. Each session of intensive vaginal dilatation lasted for 30 – 60 minutes; 2-3 times per day. Patients were able to independently perform self vaginal dilatation under direct supervision by the third day. Each patient was able to successfully attain a vaginal length of 4-5 cm with overall mean vaginal length prior to discharge of 4.62 cm. Initial pain score was 6- 8 /10, which was markedly reduced with aid of adjuvant therapy to 0-2 /10. One patient developed local vaginal tissue oedema at the entry point necessitating use of cold compress prior to dilatation therapy. No other complications were encountered. Three patients had painless menstruation following hospital discharge. All of them were continuously performing self-dilatation at home as taught and clinic review in two weeks' time following discharge showed progressive increase in vaginal length to 5-6 cm. Limitations encountered during this therapy were mainly costs of dilators and the hospital admissions.

However, these were unavoidable as prior self–home dilatation sessions were unsuccessful. We are currently looking into providing out-patient daycare intensive vaginal dilatation services.

In conclusion, non-invasive increment of vaginal length is achievable via intensive vaginal dilatation therapy. This is a promising approach even among Asian adolescents and young adults with various religious and cultural limitations. A dedicated team coupled with clear, directed and patient-centred protocol with usage of adjuvant therapy were all found to contribute to successful outcomes. Self-motivation and drive to continue self-dilatation and sound understanding of disease pathology and sequelae were important pushing factors to encourage good compliance among the patients.

After each intensive dilation session, the vaginal length is recorded. This may help boost the patient's motivation to continue with the sessions.

Figure 1. Hand-held mirror, dilators of different sizes and rulers.

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UTERINE FIBROID IN A PATIENT WITH MRKH SYNDROME

Dr Ana is a Consultant in Obstetrics & Gynaecology in Sabah Women & Children's Hospital (HWKKS). She had undergone her training in fertility preservation especially in ovarian tissue cryopreservation at the Institute for Experimental and Clinical Research (IREC) Université Catholique de Louvain, Belgium for one year. She is a Fellow of the International Federation of Paediatric and Adolescent Gynaecology (IFEPAG) in Melbourne, Australia (2019) and the first (and only) Paediatric and Adolescent gynaecologist in Borneo.

Case summary

A 44-year-old female, who was a known case of Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome, presented with complaints of intermittent lower abdominal pain with constipation for 3 months. At the age of 20, she was diagnosed with MRKH Syndrome. Her karyotype was normal (46XX). She got married at the age of 34 years. There was no complaint of difficulty during sexual intercourse with her partner.

On examination, she is phenotypically female, with normal BMI. Abdominal examination revealed a 16 weeks size mass on palpation, non-tender, firm to hard in consistency. It measured approximately 8 x 8cm and was mobile. There were no ascites or hepatosplenomegaly. The breasts and pubic hair were in Tanner stage 5 with normal female external genitalia. Vagina examination revealed a good length of vaginal canal of 2 cm and cervix was not palpable. On rectal examination, lower border of the pelvic mass could be felt.

Transabdominal ultrasonography showed a hypoechoic, heterogenous central pelvic mass of 7 × 8 cm. The kidneys were in normal in site, size, and position. CT scan of abdomen showed a well encapsulated mass in the midline of pelvis 10.2 × 8.7 × 9.3 cm as shown in Figures 1-3. Uterus could not be seen and right ovary was normal. Normal left ovary was not visualized. Kidneys are normal. Laboratory investigations were all normal. Tumour markers (Ca125, CEA, AFP, LDH) were normal.

Our differential diagnoses were of uterine fibroid (arising from uterine bud) or left ovarian fibroma. The patient was subjected to a laparoscopic intervention, during which we found an 8 x 8cm midline leiomyoma arising from the uterine bud (Figures 4-5). Both ovaries and fallopian tubes were normal. After identification of both ureters, the tumour was removed laparoscopically. Histopathological evaluation confirmed a leiomyoma without a malignant component. The patient had no surgical or clinical complication and was discharged well a day after surgery.

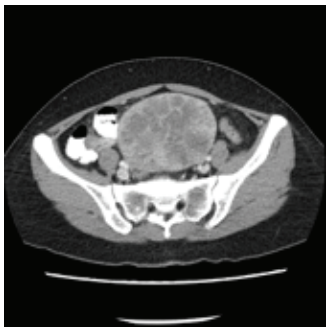

Figure 1

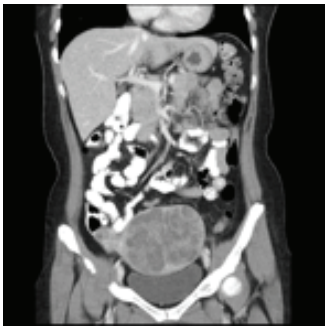

Figure 2

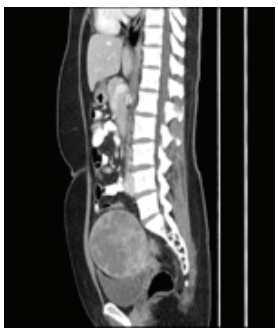

Figure 3

Discussion

MRKH syndrome, also known as Mullerian agenesis is a rare congenital disorder resulting in various degrees of malformation of vagina, cervix and uterus. The typical feature is the absence of the uterus and the upper part of vagina with normal ovaries and fallopian tubes1. While uterine fibroid or leiomyoma is a common benign pelvic tumour in women, there were only few reported cases of presence of uterine fibroid in MRKH syndrome2-18. Women with MRKH syndrome have a normal 46XX karyotype, normal level of female secondary sexual characteristic and ovarian functions. Under the influence of estrogen, rudimentary uterus or proximal end of Mullerian ducts which contain smooth muscles may be stimulated and promote the estrogen dependent diseases such as leiomyoma and adenomyosis14.

Diagnostic laparoscopy played an important role as a diagnostic tool and definitive surgical intervention especially in this case as her investigations were not suggestive of cancerous lesion instead of proceeding with laparatomy9. In conclusion, uterine fibroid may arise in MRKH women and laparoscopic assessment and interventions should be considered as a standard management.

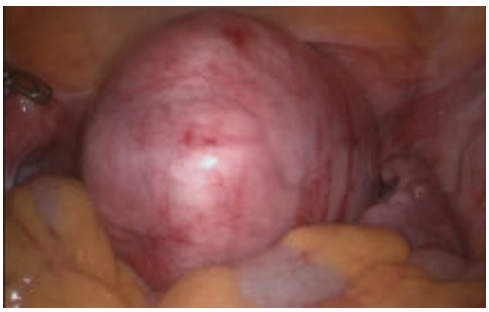

Figure 4. Uterine fibroid arising from uterine bud.

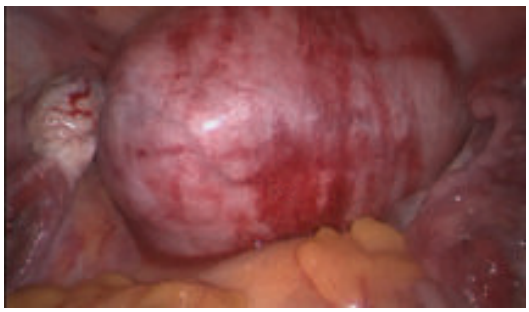

Figure 5. Uterine fibroid arising from uterine bud.

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