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醫學文章
Medical Articles

專題文章
Special Topic Article

本港公營唇顎裂治療概論
Overview of Cleft Services in Hong Kong

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唇裂顎及相關缺陷需要接受跨部門綜合治療，這包括產前診斷及父母輔導，以至唇裂及顎裂修補，及在成長過程中矯正言語、牙齒和面部外觀都十分重要。

唇和顎早在妊娠第六週就開始發育。在早期產前檢查中，可以透過超聲波和造影檢測到顏面缺陷問題。單側或雙側唇裂容易確定，但要確定顎裂比較困難。如單純只有唇顎裂的問題是不需要終止懷孕，因為可以透過手術來矯正。懷疑有唇顎裂胎兒的父母可以在生產前，透過轉介到唇顎裂診所了解治療程序和接受輔導，在照顧方法和心理上得到支援。

在本港，唇顎裂診所設於兒童醫院、瑪麗醫院、廣華醫院及屯門醫院，如唇裂顎問題是在產後才發現，病人將會被轉介到相關的診所跟進治療。大部分診所可在一、兩個星期內安排第一次見醫生，所以家長無須憂慮要等候很長時間才可見醫生。在私家治療方面，亦有數位醫生做唇顎裂手術，然而對病人來說，長時間的綜合治療會給予更好的跟進和治療效果。

現時，唇裂及顎裂會分別在 3 個月大及 9 個月大時進行修補手術。手術前，家長須學習照顧孩子，尤其是使用特別奶瓶餵哺，公立醫院在產後會提供餵奶指導，另本港亦有病人組織 - 香港兔唇裂顎協會 - 為患者家長提供支援和協助。

修補手術需在患兒達到一定年齡和體重才進行，以確保手術安全。然而，某些個案如早產嬰、患短下巴和有呼吸問題的皮爾羅賓症患兒，又或是患有其他疾病以至在全身麻醉期間和手術後護理構成風險等，將會延遲修補手術。

手術前，可配戴鼻顎托作矯正寬闊裂隙、外突前牙骨、鼻裂等問題。至於較為輕微的個案，鼻顎托的使

The management of cleft lip and palate and associated condition is a multidisciplinary care. It starts from antenatal screening and counselling of parents to surgical correction of the cleft lip and palate anomalies, to be followed by correction of speech, dental and facial appearance throughout the growth development of the affected cleft patients.

Normal lips and palate develop as early as 6 weeks of gestation and the presence of clefting can be detected with modern ultrasound and imaging during antenatal care in the first trimester. Cleft lip, isolated or bilateral, can be identified whereas cleft palate may sometimes be difficult. For isolated cleft conditions, termination of pregnancy is not necessary as cleft is a surgically correctable condition. Parents with suspected cleft fetus can be referred to cleft clinic for counselling and explanation of the management protocol well before delivery. This provides information on care and psychological support of parents with the cleft condition.

In Hong Kong, cleft clinic is located in Hong Kong Children's Hospital, Queen Mary Hospital, Kwong Wah Hospital and Tuen Mun Hospital. If the detection of cleft is only identified after delivery, patient can also be referred to the respective clinic for further management. Most clinic will arrange the first appointment within 1 to 2 weeks without the worry of long waiting time. In the private sector, a few surgeons will do cleft operation but a more extended multidisciplinary care is crucial to give the best outcomes.

Currently cleft lip and palate are surgically corrected at 3 months and 9 months respectively. Before surgery, parents will be educated on the care of cleft baby, particularly on feeding and the use of special feeding bottles if indicated. Nursing advice is available in hospitals after delivery. A patient group, the Hong Kong Association for Cleft Lip & Palate, also provides support and advice to involved parents.

Surgery is performed when the child attains certain age and body weight to allow safe surgery. Delay in surgical repair may be indicated in selected

用仍具有爭議性。配戴鼻顎托，家長必須堅持，而膠布貼在患兒面上亦可能引起皮膚敏感，須特別留心。

唇裂及顎裂的修補手術會由整形外科或小兒外科醫生提供，同時得到其他專科如耳鼻喉科、牙科等配合，每個診所都有自己的治療模式去確保照顧和治療效果，亦會跟進在私家或海外接受手術的病人。唇顎裂治療由出生開始至成人階段才完成，療程較為漫長。

在一歲前，外科醫生會完成唇裂及顎裂的修補，及後言語治療師會評估患兒的語言發展，並由兩歲開始提供適當治療，包括咬字、發音和吞嚥訓練。由於現時已經有駐學校言語治療服務，這可以分擔公營言語治療服務的壓力，為有較嚴重言語問題的患童提供治療。

牙科外科醫生會在不同的成長階段中跟進患童的牙齒生長，提供牙床植骨手術，及顎骨矯正手術以改善上下牙齒倒崁問題，亦會在適當時間轉介患者接受矯齒治療。

患有嚴重顎裂，尤其雙側唇顎裂患者，他們的前齒骨及牙床骨發育較差，導致細少、收縮及三角形上牙槽。上牙槽前方細小和狹窄，亦可致牙齒擠迫和錯位。上下頜生長的差異會導致牙齒咬合不正常。唇顎裂患者普遍有上頜後縮和下頜前突，或兩者兼有的第三類不正常牙齒咬合。此外，後退的上頜骨會在面部輪廓上產生面部中央凹陷，這是典型的扁平臉頰外觀。牙齒矯正治療有助擴張上牙槽，並重新排列牙齒。而正顎手術會推出上頜骨，改善面部輪廓和上下牙齒的咬合。這種牙骨矯正手術通常會在 16 至 18 歲面部發育完成後才進行。

在 2014 年之前，針對狹窄上顎和牙齒錯位的矯齒治療在公營醫院服務有限。現時，矯齒服務由醫院管理局提供。由於資源有限，醫管局只聘請一位矯齒科醫生，長駐在聯合醫院牙科部，所有唇顎裂中心都可以轉介病人去接受矯齒治療。這確保了對唇裂和顎裂患者的持續治療，並有助於日後進行正顎手術，改善牙齒咬合和面部輪廓。

此外，耳鼻喉外科醫生會負責跟進患者的中耳積液和放置引流管，兒科醫生會負責跟進患有唇顎裂相關綜合症的問題，臨床心理學家則會為有需要的父母和患

situation such as preterm delivery, Pierre Robin Sequence with small jaw and anticipated airway problems, or other premorbid medical conditions that make the anaesthesia and postoperative care at risk.

The use of pre-operative orthodontic care, the nasoalveolar molding (NAM) is getting more popular. While it is commonly practiced in wide-gapping cleft, significant premaxilla protrusion and alar flaring, the use in less severe cleft is still controversial. It requires good compliance of parents and may cause irritation on the child's face with the adhesive taping.

Surgical repairs of cleft lip and palate are performed by a team of surgeons, be it Plastic or Paediatric surgeons, and supported by other disciplines e.g. ENT surgeons, dental surgeons. Each centre will has its own protocol to ensure good quality of care and associated treatment. They will also provide continued care of patients with operation done in private or other places overseas. In fact, the patient care journey is a long run up to adulthood.

While the surgeons will complete the surgical correction of cleft lip and palate usually before one year of age, the speech therapists will assess the speech development and provide appropriate training as needed from 2 years onwards. These include articulation, phonation and swallowing training. With the support of school based speech therapists in most setting, it helps to diversify the workload to allow more resources and concentration on complicated patients.

The dental surgeons will be involved in monitoring the growth of dentition, advice and care on dental caries, restoration of the upper alveolar arch by performing alveolar bone grating (ABG) in presence of alveolar bone clefting. They will also provide orthodontic and orthognathic treatment. The timing of treatment depends very much on the various phases of care required as the child grows with time.

In patients with severe cleft palate and particularly in bilateral involvement, the maxilla and alveolar bone may be underdeveloped leading to small, contracted and triangular arch of alveolar. The limited space leads to crowded teeth with displacement and entrapment. Discrepancy of growth of upper and lower jaw will lead to malocclusion. Class III malocclusion with maxillary retrusion and mandibular protrusion or a combination is more common in cleft patients. Moreover, the receding maxilla will create a central depression on the facial profile, a typical flat cheek appearance. Orthodontic care helps to expand the contracted alveolar and realign the teeth while orthognathic surgery will recreate the normal facial profile and occlusion of upper and lower jaw by cutting and advancing the maxilla bone after

者提供心理輔導，以上都是綜合治療所提供的治療重要方案。

在唇顎裂第一期手術後，接著是第二期唇顎裂手術，包括唇部和鼻部整形手術，改善患者的唇鼻結構和軟組織，使療程更為完整和完善。第二期唇部和鼻部整形手術包括：疤痕改善、唇部軟組織增加、口鼻漏管消除、改善說話鼻音過重或顎咽閉合不全的矯正等。鼻部整形有助改善鼻部外觀欠佳，因應鼻部缺陷的嚴重程度來取決需否自體骨移植，以達至較美觀的手術效果。

唇部和鼻部整形手術的時間視乎要考慮患者的面容差異，疤痕預期收縮程度和成年後唇鼻外形變化的平衡。唇顎裂治療不限於嬰兒階段，也沒有年齡限制。醫生會跟進患者個案一直至成年期，以確保達至功能和美觀效果。公營醫院的整形及整容外科部亦接受成人唇顎裂患者和其他顱顏缺陷病人尋求治療。

隨著新發展，唇顎裂治療仍不斷在演變。經改良的手術方法可使手術效果更為理想。在裂唇修復時，使用肉毒桿菌注射和早期激光治療改善唇部疤痕，有助於減少唇部拉張和改善疤痕收縮。如不能廣泛採用於所有患者，至少可以為有需要的患者提供，這仍需要更多的研究來證明這種輔助治療的有效性和好處。

唇顎裂治療不是單一項的手術，需要在不同的生長階段為患者提供跨部門的綜合治療，持續跟進是至關重要。

completion of orthodontic treatment. This bone work will usually be carried after completion of facial bone growth at the age of 16 to 18 years of age.

Before 2014, orthodontic care for contracted palate and associated dental misalignment is limited in the public sector. Nowadays, orthodontic care is supported by Hospital Authority. Due to resource implication, orthodontic care is centralized within HA and is located in United Christian Hospital dental unit. All cleft centres can refer cases for orthodontic need. This ensures a continued care of cleft lip and palate and facilitates subsequent orthognathic correction of malocclusion and facial profile.

Besides, other parties such as ENT surgeons for middle ear effusion and myringotomy tube insertion, Pediatricians for management of associated syndromic conditions, clinical psychologists for clinical support to parents and patients etc. are equally essential in the multi-disciplinary approach.

Following primary correction of cleft lip and palate, secondary cleft surgery and associated rhinoplasty correction provide refinement of both structural and soft tissue reconstruction, making a comprehensive treatment of the cleft and associated problems. Examples of secondary procedures include scar revision, tissue bulk augmentation, fistula correction, velopharyngeal insufficiency correction for hypernasality and others. Rhinoplasty with or without cartilage or bone graft also help to improve the aesthetic appearance of associated cleft nose deformities. The timing of repair depends on a balance of the severity of disfigurement, anticipated scar contracture during facial growth and further plan for structural correction. Cleft management is not limited to infancy and there is no age limit. Patient follow up extends towards adulthood to ensure both functional and aesthetic reconstruction are achieved. Adult patients with cleft lip and palate problems are accepted and patients with other craniofacial anomalies will also be managed.

With recent advances, the treatment of cleft is still evolving. Modification of surgical techniques improves the surgical outcomes making the scar and reconstruction more pleasing. The use of botox injection during lip repair and early vascular laser on lip scarring helps to minimize tension and control early maturation of scar. This can be offered to high risk group if not to all patients. More studies are required to prove the efficacy and advantages of such ancillary treatment.

Cleft management is not one single surgery and a multidisciplinary, appropriate care at different timeline of growth is essential. A continued care should be provided.

醫學文章

整形外科在顱顏中心的角色
Role of Plastic Surgery in a Craniofacial Centre

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顱顏手術是治療頭顱和顏面的骨骼和軟組織的缺陷和異常，包括單獨性的缺陷以至不同嚴重程度的其他顱顏畸形，並可能涉及症群，影響聽力、呼吸、吞嚥和智力發展。

對顱顏缺陷的患者來說，跨專科綜合治療是最理想的治療模式，可以提供全面的和經協調的治療計劃。

在任何顱顏中心，都應該有一個專家團隊，包括整形和整容外科醫生、神經外科醫生、口腔頜面外科醫生、小兒外科醫生、矯齒科醫生、言語治療師、聽力專家、兒科醫生和物理治療師。

整形和整容外科是一門獨特的專業，負責重建、功能改善，並顧及正常外貌和美觀為原則。然而，整形及整容外科可能是最容易被誤解的外科專業之一，原因是一般人以為整形及整容外科主要處理美容手術，其實，重建手術才是整形外科醫生的主要工作，是驅使我們不斷求進的動力。

整形及整容外科當然在顱顏缺陷治療擔任重要的角色。

先天性缺陷

唇顎裂是最常見的先天性顱顏缺陷。其他較為少見的顱顏缺陷包括顱縫早閉、半邊小臉症、耳朵畸形、綜合畸形症狀，如特雷徹柯林斯綜合症等等。

當中一些手術會根據孩子的發育情況，分階段進行，因此長期和持續的跨專科的治療對於這些患者非常重要。

Craniofacial surgery treats conditions that affect bones and soft tissues of the head and face. Problems can range from isolated defects to multiple malformations of various severities, and can be associated with deficiency in hearing, breathing, swallowing and mental development.

Multidisciplinary approach is definitely the gold standard with the aim of delivering a comprehensive and coordinated treatment plan for patients with craniofacial deformities.

In any craniofacial centre, there should be a team of specialists including plastic surgeons, neurosurgeons, oral & maxillofacial surgeons, pediatric surgeons, orthodontists, speech therapists, audiologists, pediatricians and physiotherapists.

Plastic and reconstructive surgery is a unique specialty which involves the concept of reconstruction, functional and aesthetic enhancement. However, it is probably one of the most misunderstood surgical specialty. The general misconception is that plastic surgeons only deal with aesthetic issues. However, reconstructive surgery is in fact the main focus and driving development in plastic surgery.

Plastic surgeons certainly can have an active role in management of craniofacial deformities.

Congenital deformities

Cleft lip and palate are by far the commonest congenital craniofacial problems. Less common conditions include craniosynostosis, hemifacial microsomia, ear anomalies, syndromic deformities e.g. Treacher Collins.

Some of these surgeries are staged according to the child's development, therefore long term and continuous multidisciplinary care is very important for this group of patients.

意外性創傷

不同年齡組別的意外性創傷病人需要顱顏重建。很多時候，重建手術需要分階段進行以獲得形態和功能上的全面復原。

腫瘤和癌症

神經纖維瘤是一種使人沮喪的問題，通常需要進行多階段的切除和重建手術。當神經纖維瘤涉及面部時，手術尤其具有挑戰性。

面部和顱骨的腫瘤（不管是良性或者惡性），在切除後，需要進行複雜的重建手術。在這些情況下，通常會以跨部門團隊形式提供治療。

面部活動機能恢復

有些患有先天性面部缺陷涉的病人面部神經功能有障礙，如半邊小臉症、牟比士症候群，面部神經創傷性受損導致面癱，這些患者都可以從面部活動機能恢復手術中得到改善。

血管異常

面部和頸部血管異常可能發生在兒童和成人身上。治療通常以跨專科形式進行治療，團隊成員包括介入性放射治療員、整形外科醫生和麻醉師。如同時需要進行手術切除，通常由整形外科醫生執行。

燒傷

面部的嚴重燒傷個案通常會在燒傷中心進行處理。然而，第二期頭顱和面部的缺損可以在顱顏中心進行。慢性燒傷疤痕可使人感到沮喪，尤其疤痕位於眼簾上方或口部會影響眼部和嘴巴打開。大多數這些疤痕需要一次性或多階段的重建。

總結而言，整形及整容外科醫生在顱顏缺陷治療發揮著至關重要的角色，尤其在身體器官重建方面和長遠的形態修復和外觀改善。故此，整形及整容外科絕對是每一個完善的顱顏中心跨專科團隊的成員之一。

Trauma

Patients of all ages with trauma-related injuries may require craniofacial restoration. Very often, staged procedures are needed in order for a full restoration of function and form.

Tumors and cancers

Neurofibromatosis is a debilitating disease which normally requires multi-staged surgeries involving resection and reconstruction. Facial involvement is particularly challenging.

There are also benign and malignant tumors involving the face and skull which require complex reconstructions after tumor resection. Combined team approach will usually be adopted in these conditions.

Facial reanimation

There are congenital conditions where facial nerve does not function, e.g hemifacial microsomia, moebius syndrome, facial nerve injuries in trauma can also result in facial palsy. These patients can benefit from facial reanimation procedures.

Vascular anomalies

Vascular anomalies involving the face and neck can occur in both children and adults. They are usually managed in a multidisciplinary manner involving interventional radiologists, plastic surgeons and anaesthetists. There are times when surgical excision is needed and will usually be performed by plastic surgeons.

Burns

Acute burns involving facial area will usually be managed in a burn centre. However, secondary deformities involving the skull and face can be managed in a craniofacial centre. Chronic burn scars can be debilitating, especially if there are located over the eyelids or restriction mouth opening. Most of these scars will likely need single-staged or multi-staged reconstructions.

In summary, plastic surgeons play a crucial role in craniofacial conditions especially the reconstructive aspect, although aesthetic adjustments are often needed in the long run. Plastic surgery should definitely be part of the multidisciplinary team in a well-developed craniofacial centre.

醫學文章

單側唇裂及巨口症修補手術
Surgical Repairs for Unilateral Cleft Lip and Macrostomia

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簡介

唇顎裂是可以透過手術矯正的。儘管如此，患兒和及家長在孩子整個成長發展過程中都會面對各種挑戰。在現代的唇顎裂治療中，患兒會由有經驗的醫療團隊照顧，包括來自不同專業的外科醫生、牙醫、牙齒矯正科醫生、言語治療師、護士、物理和職業治療師，團隊甚至在嬰兒出生前就已開始計劃裂唇治療。在產前超聲波檢查（USG）診斷確定胎兒患有唇顎裂後，父母應接受外科醫生的輔導，讓家長能夠事先為即將面對的挑戰作好生理和心理上的準備。

手術的時序

一般來說，患兒的第一次手術是裂唇修補，這是一項全身麻醉的手術。雖然理論上裂唇修補手術可以在任何年齡進行，但在平衡手術的風險和好處之下， 手術通常會在患兒約 3 - 6 個月大進行，且會按患兒裂唇的程度和身體狀況而訂定，修補時間和手術的方法也會各院有異。

手術前的準備（鼻顎托 NAM / 上唇貼）

患兒出生後，應盡快開始貼唇貼。那些上唇裂隙較寬的患兒在補唇手術前配戴一段時間的鼻顎托 (NAM) 可能會有助於改善牙槽、減少唇裂寬度和鼻子塑形。然而，鼻顎托需要由牙科醫生提供，並且在整個治療期間也需要緊密的跟進和父母的全程投入。因此，在某些公立醫院可能無法進行，並且非所有家庭都能在整個治療中配合。除了鼻顎托外，還可以為唇裂患兒貼上唇貼，以收窄上唇裂隙，有助將來的唇裂修補手術。

Introduction

Cleft conditions are surgically correctable, nevertheless, babies with cleft lip or facial cleft and their families will face various challenges throughout the growth and development of the children. In modern cleft services, children with cleft conditions are looked after by a team of experienced health care providers including surgeons from different specialties, dentists, orthodontists, speech therapists, nurses, physio- and occupational therapists. Management of cleft condition starts even before the baby is born. Parents should receive counselling by cleft surgeons once the diagnosis is made by antenatal USG screening. The family can then be better prepared for the upcoming challenges physically and also mentally.

Timing of operation

Very often, the first surgery that cleft babies will encounter is the lip repair. Lip repair is a general anaesthetic procedure. Although cleft lip repair theoretically can be performed at any age, usually it is done at around 3 to 6 months of age balancing between the risk and benefit of the operation. The timing of repair and the operative technique vary among institutions and will be tailor made to accommodate the phenotype of the cleft and the babies' other medical conditions.

Presurgical management (NAM/ lip adhesion)

After the baby is born, lip taping should be commenced as soon as possible. For those wider cleft lips, babies may benefit from a period of nasoalveolar molding (NAM) before definitive surgical repair in terms of molding the alveolus, decreasing cleft width and with the advantage of molding the nose. However, NAM treatment requires expertise and also intensive follow up care and parents' input throughout the treatment period. Therefore, it may be unavailable in some institutions and not all families can be compliant throughout the whole treatment. Apart from NAM, lip adhesion (labial adhesion) might be offered to selected cleft lip babies in order to facilitate future definitive lip repair.

單側唇裂修補

自 1900 年代以來，單側唇裂修補手術的技術已經從直線修復發展到現今最流行的兩種修補技術 — 旋轉推進法和幾何式修復。這兩種修補技術在有經驗的外科醫生操刀下能有理想的手術效果；但同時這兩種技術亦有其不足之處。 米勒德 (Millard) 推出了旋轉推進修復技術，這技術現今已成為最流行的修復技術之一。透過這種技術，可以塑造一個對稱的，如丘比特弓的上唇，而裂隙修補的疤痕非常類似人中柱，使患兒在患側和健側上唇都有人中柱。隨著時間的發展，不少外科醫生改良這種修復技術。現時，世界各國最廣泛採用的是莫勒 (Mohler) 的修復技術，其中裂隙修補的疤痕更垂直，更像真天然的人中柱。然而，這些修改方法通常會產生更複雜的疤痕，尤其是在人中柱與上唇的交界處，並且需對裂唇的水平長度作出妥調。此外，旋轉推進修復技術使用了一種“隨取隨用”的技術，這對較年輕的外科醫生來說可能是比較難以掌握。

三角瓣修復術 (Tennison 和 LeMesurier) 是幾何式修復技術的先驅。然而，這技術所產生的疤痕會破壞人中柱的完整性，所以受到批評。 費希爾 (Fisher) 發表了他對幾何式修復技術的改進修復術。這技術所產生的疤痕能準確地吻合了健側人中柱，而鼻樑和鼻翼周圍亦沒有額外的疤痕。三角瓣剛好在丘比特弓正上方，這不僅加長了上唇長度，上唇唇弓更加對稱，亦避免垂直式疤痕所導致的強烈收縮問題，所以這手術方法獲唇顎裂外科醫生普遍採用。

在旋轉推進和幾何式修復技術中，唇紅高度的不足可以通過三角瓣從健側唇紅線處來補足。這可以避免為使用橫向紅唇組織而導致上唇有吹口哨動作的困難。

上面討論的技術僅與皮膚切開術有關。對於成功的裂唇修復，肌肉和黏膜修復也非常重要，可以顯著影響最終修復的效果。良好的裂唇修復可使上唇環狀肌肉修復，使上唇活動自如，避免上唇有吹口哨動作的困難，並有助於矯正鼻畸形。唇裂和鼻畸形也可以在唇部修復手術中同時進行。鼻修復的目的是為鼻子打下良好基礎有助以後生長。

患側鼻翼缺陷亦可以在裂唇修補同時進行。在裂唇修復時，可以打開患側鼻翼邊緣或在唇部三角瓣切口處加額外切口，矯正鼻翼軟骨後再縫合。

Unilateral cleft lip repair

The surgical technique of unilateral cleft lip repair has evolved since 1900s from a straight-line repair to the two most popular designs nowadays – rotation-advancement method and geometric-style repair. Both techniques, when in experienced hands, can provide excellent result; but at the same time, each of them has its own intrinsic insufficiency.

Millard introduced the rotation-advancement technique which became one of the most popular repair techniques nowadays. With this method, a balance Cupid's bow can be created with the resultant scar closely resembles a continuous philtral column. With time, this technique was refined by different surgeons and the most widely adopted modification world wide is Mohler's modification in which the incision lies more vertically to better correspond to the natural philtral column. However, these modifications usually incorporate more complex scars especially around the columellar-labial junction and also can compromise the cleft side horizontal lip length. Also, rotation-advancement technique utilized a "cut-as-you-go" technique which young surgeons might find it more difficult to master.

Tennison and LeMesurier type of repair was the pioneer in geometric-style repair. These techniques were critized by the disruption of continuity of the philtral column by the resultant scar. Fisher published his anatomical subunit approximation technique which is evolved from the earlier geometric-style repair. With his technique, the resultant scar exactly mirrors the contralateral philtral column with no additional scar around the columella and alar. The triangular flap just above the Cupid's bow not only provides additional vertical lip length for a balance Cupid's bow, it also breaks up the linear scar to prevent scar contracture. And this technique is gaining popularity among cleft surgeons.

In both rotation-advancement and geometric-style technique, deficiency in vermilion height can be overcome with a triangular flap from lateral lip at the red line. This can prevent whistle deformity by utilising tissue from lateral lip segment that would have been discarded anyway.

The technique discussed above is about the skin incisions only. For a successful cleft lip repair, muscle and mucosal repair is also very important and can affect the final outcome significantly. A good orbicularis oris muscle repair provides oral competence, prevents whistle deformity and helps to correct nasal deformity as well.

Cleft lip nasal deformity can also be corrected simultaneously during lip repair operation. The aim of repair is to establish a good foundation for future growth of the nose. Cleft side lower lateral cartilage

巨口症的修復

巨口症是一種罕見的先天性缺憾，也是一種顏面缺裂，分為單側性和雙側性缺裂。裂隙涉及皮膚、肌肉和黏膜層。巨口症修復旨在重建口腔活動能力（功能性），同時要使唇部和口部對稱（外觀性）。文獻中論述了不同的巨口症修復手術技術。原則上來說，黏膜、口輪匝肌和皮膚需分三層縫合，並且通常採用 Z 形手術技巧，以取代簡單直線縫合所造成的嚴重疤痕收縮問題。

結論

像所有其他整形外科手術一樣，裂唇和巨口症修復是一項為個別病人特別制定的手術。無論外科醫生採用那種修復技術，修復方法都會有所調整，以針對不同類別的裂唇狀況。因此，很難直接比較不同技術的手術結果。儘管如此，我們都有一個共同目標 - 就是要達到對稱和平衡的上唇，讓唇裂患兒能夠在身心健康的情況下茁壯成長。

can be freed via existing lip incision or additional incision at the soft triangle and suspended with sutures.

Macrostomia repair

Macrostomia is a rare congenital condition and is a type of facial cleft, it can be unilateral or bilateral. The cleft involves the skin, muscle and mucosal layer. Macrostomia reconstruction is aiming to reconstruct the oral competence (functional) and to obtain a symmetrical lip and commissure (cosmetic). There are different surgical techniques described in literature for macrostomia repair. In principle, mucosa, orbicularis oris and skin are closed in 3 layers. And often multiple Z plasty are incorporated in the skin closure to break up a long linear scar and to prevent future scar contracture.

Conclusion

Like all other plastic surgeries, cleft lip repair or macrostomia repair is an individualised operation. No matter which technique the surgeon adopted, there will be variation among different cleft repairs in order to cater different phenotypes of the defect. Thus, it is very difficult to directly compare outcomes of different techniques. Nevertheless, we all have a common goal, to obtain a symmetrical and balanced lip so children with cleft conditions can thrive with good health mentally and physically.

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雙側唇裂修補及鼻整形手術
Bilateral Cleft Lip Repairs and Nose Revision

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介紹

雙側唇裂是一種先天性的缺陷，其中嬰兒的上唇兩側不相連，其範圍可以從唇邊的小裂縫至局部範圍，甚至整個唇部。香港每 500 名新生兒中約有 1 人患有唇裂，其中約十分之一患有雙側唇裂。

雙側唇裂有時會伴有顎裂，鼻子和上牙齦可能會出現相關畸形， 百份之三十 的嬰兒也可能由某些綜合症候群引致其他畸形。如果沒有確診綜合症候群，在家庭中遺傳的機率相對較低（約 5-15%），這取決於有多少家庭成員患有此病。母親年齡 大於 40 歲是一個危險因素。懷孕期間吸煙和飲酒是最常見的環境因素。

雙側唇裂的問題 / 畸形

對雙側唇裂嬰兒主要的影響是外觀。如果他們沒有伴隨顎裂，縱使餵食可能會受到影響，但他們通常仍然可以通過母乳或奶瓶餵食。

畸形情況大致如下：

The deformity would be as following:

- 上唇兩側唇部不相連（隱性 / 不完整 / 完整的唇裂）可能對稱或不對稱
  Discontinuity of lip on both side of upper lips (Microform/Incomplete/Complete), which may or may not be symmetrical
- 唇紅緣（唇中央部份全厚度的軟組織，包括皮膚、軟組織和黏膜）直接掛在鼻尖皮膚上
  Prolabium (The central segment of the full thickness soft tissue lip including skin, soft tissue and mucosa) hang directly from the nasal tip skin
- 前上頜骨突出（上頜牙弓中央骨部份）
  Protrusion of premaxilla (The central bony segment of maxilla at dental arch)
- 雙側鼻翼塌陷，短鼻樑，鼻頭形狀扁平
  Bilateral nasal alar collapse, short columella and broad flat nasal tip
- 前鼻棘形成不良或缺失，導致鼻中隔軟骨底部後移和內側腳底板緊
  Poorly formed or absent anterior nasal spine, resulting in a retruded area under the base of the septal cartilage and recession of the foot plates of the medial crura

Introduction

Bilateral Cleft lip is a birth defect in which a baby's upper lip is in discontinuity on both sides of the lip, which can range from small notches at the edge, to involvement of partly or the whole height of the lip. Around 1 in 500 newborns in Hong Kong has cleft lip, and around one-tenth of them has bilateral cleft lip.

Bilateral cleft lip can sometimes come with cleft palate. There may be associated deformity of the nose and upper gum. Thirty percent of the babies may also have other deformities explainable by certain syndromes. If there is no diagnosed syndrome, the chance of it running in the family is relatively low (~5-15%), depending on how many of the family members has the condition. Age of mother >40 years old is a risk factor. Smoking and alcohol use during pregnancy are most common environmental factors.

Problems/deformity in bilateral cleft lips

The major impact on babies with bilateral cleft lip will be on its appearance. If they do not have associated cleft palate, although feeding may be affected, they can usually still be fed via breast-feeding or bottle feeding.

治療流程
Protocol

新生兒 Newborn	鼻槽成型療法 nasoalveolar molding
3 個月 3 months	裂唇修復手術 Lip repair
9-18 個月 9-18 months	裂顎修補手術（如伴有顎裂） Palate repair if associated with cleft palate
4-6 歲 4-6 years	如有需要時，進行顎咽閉合不良手術（顎裂病例） Surgery for velopharyngeal insufficiency if necessary (cleft palate cases)
6-18 歲 6-18 years	牙齒矯正。在 9-12 歲左右進行牙床植骨手術。如有需要，隨後會進行正顎手術。 Orthodontic treatment. Alveolar bone graft at around 9-12 years old. Orthognathic Surgery afterwards if necessary.
16-18 歲以後 16-18 years onwards	如有需要時，進行裂鼻外觀整型手術和唇成形手術 Cleft Rhinoplasty and lip revision if necessary

餵食

在母乳喂食期間，乳房通常會被嬰兒嘴唇 / 牙齦的開口覆蓋。唇裂可能影響嬰兒不能在乳頭周圍形成完全的包封，用手指蓋住裂隙可能會有幫助。而對於用奶瓶餵食的，可能用底部較寬的人工奶咀為佳。

術前頜面矯形學操作 -- 鼻顎托

新生嬰兒在特定時期，他們的畸形是具有“可塑造伸延性”的。因為他們的骨骼和軟組織具有高度的可塑性。這可能是由於其細胞間基質中的透明質酸含量較高。

鼻顎托有 3 個組成部分：
1. 鼻構形器
2. 上顎托
3. 上唇貼

這 3 個部份可以組合在一起或分階段進行，可先進行牙槽矯形再作鼻矯形，鼻顎托可配戴至裂唇修復前，惟家長必需配合頻密的覆診，每星期帶患兒見牙科醫生以調整鼻顎托。

配戴鼻顎托的目的是使雙側唇裂患兒的前上牙槽骨回縮，同時矯正鼻翼軟骨和鼻樑畸形。術前配戴鼻顎托可減少裂唇和鼻的畸形，減低修復手術的困難程度，亦可減少患兒早期快速成長過程中出現的鼻部變形。

Feeding

Cleft lip may affect the baby forming a good seal around the nipple. During breast-feeding, the breast often will fill the opening in lip/gum. It may be useful to hold a finger across the cleft. For bottle-feeding, artificial teats with wider base may be useful.

Presurgical dentofacial orthopedic manipulation--Nasoalveolar molding

There is a special period of time for a newborn to have their deformity ‘moldable’ because their skeleton and soft tissues have a high` degree of plasticity. This may be due to a higher level of hyaluronic acids in their intercellular matrix.

There are 3 components of nasoalveolar molding:
1. Nasal conformer
2. Intraoral palate molding plate
3. Lip/cheek taping

The 3 components can come together or can be staged, with alveolar molding before nasal molding. The same plate may be used throughout the period, with modifications as frequent as weekly to sequentially modify the form of the deformities.

The goal of presurgical nasoalveolar molding in bilateral clefts is to retract the premaxilla, while at the same time achieving correction of the nasal cartilage and columella deformity. The presurgical reduction in hard and soft tissue cleft deformity reduces the magnitude of surgical challenge as well as minimizing the nasolabial distortions that occur during the rapid growth of early childhood.

第一期雙側唇裂修補手術的技術

手術將於患兒大約 3 個月大時在全身麻醉下進行。如患兒體重較輕，可能需要推遲手術時間。手術效果不會因延誤而受到影響。

雙側唇裂修復手術會利用所有上唇中間部份的軟組織。對於中央段的裂唇，外科醫生會利用前唇的皮膚加長鼻小柱外皮，而前唇黏膜將會建構前唇的表面，並加深上唇中間部份的牙齦溝。補唇疤痕會特地在人中脊位置以模仿人中脊線。
雙側裂唇的側唇部份會被切開並置於中線，將異常附著的唇肌分割，以修補上唇中線使唇部肌肉相連。保留側唇部份的黏膜，以重建上唇結節（最期待的紅唇緣黏膜在中線）和鼻孔底部。

鼻孔邊緣可能會有切口以露出兩邊鼻翼軟骨，並將它們從異常覆蓋皮膚上的部份釋放出，通過縫合技術矯正變形和錯位的軟骨。

在唇裂和顎裂的第一期修復時，上牙槽的裂隙不會修補。當孩子長大，並在進行牙槽骨移植時，上牙槽前方的口鼻漏管才會被封閉。

特殊情況

不對稱雙側唇裂

如前所述，雙側唇裂會出現一系列不同嚴重程度缺裂，大多數是不對稱的。如果雙側唇裂明顯不對稱，並且無法通過術前鼻顎托去減低，有時可能需要分階段進行重建程序以達到最終最佳成效。

現有結構發育不足

一些嬰兒可能患有某些異於常見類型的雙側唇裂，他們可能有發育不良的中線結構。他們可能鼻中隔過短，鼻樑前方組織不足， 鼻小柱呈圓錐形，前唇和上牙槽前齒骨發育不全，上牙槽前齒骨下垂、上唇紅唇部份很薄，使術前鼻顎托亦無濟於事。修補此類唇裂畸形的技術可能需要修訂，第一次修補的結果可能不夠理想。這些兒童可能需要第二次修整手術，包括添加自體真皮脂肪和軟骨移植。

純粹雙側唇裂而並無顎裂

無顎裂的純粹雙側唇裂並不常見。但如有這情況，上牙槽前齒骨可能比較堅實，使術前鼻顎托無效。如上牙槽前齒骨妨礙裂唇修補，則需要先將其矯正。如上顎完好無損，中面生長的負面效果不會因為上牙槽前齒骨的矯正而受到影響。

Surgical technique for primary bilateral cleft lip repair

The surgery is to be done under general anesthesia at around 3 months of age. For babies who has slower weight gain, it may be necessary to postpone the surgery. Outcome of the surgery would not be affected by the delay.

Repair of bilateral cleft lip makes use of all the available local tissues. For the central segment of the cleft lip, skin of the prolabium is used for reconstruction of philtrum skin while mucosa of the prolabium is used to reconstruct the anterior surface of premaxilla and deepen the gingival sulcus of central lip segment. The design of the neo-philtrum skin is to place the scar at the expected philtrum columns and mimic them. Bilateral lateral lip segments are dissected and are brought to the midline, with the lip muscles detached from their abnormal insertions and repaired at midline to bring the upper lip muscle in continuity. Mucosae from lateral lip segments are preserved for reconstruction of tubercle of upper lip (the most projected vermillion mucosa at midline) and nostril floor.

There may be incisions at nostril rims to expose bilateral nose cartilages and release them from their abnormal attachment to overlying skin. The deformed and misplaced cartilages are corrected by suture technique.

The cleft at upper gum may not necessarily be closed during primary repair of the cleft lip and palate. The oronasal fistula at anterior upper gum will be closed during alveolar bone grafting when the child is getting older.

Special situations

Asymmetrical deformity

As mentioned previously, bilateral cleft lip is a spectrum of deformity of different severity, and it may not always be symmetrical. In case of significant asymmetry of the bilateral cleft deformity and which was not able to be minimized by presurgical nasoalveolar molding, in certain occasion it may be necessary to perform reconstructive procedures in stages in order to achieve the best final result.

Underdevelopment of existing structures

Some babies may suffer from certain variations of cleft deformity different from the usual type of bilateral cleft lip. They may have underdeveloped midline structures. They may have short nasal septum, absent anterior nasal spine. The columella may be in cone shape. Prolabium and premaxilla are underdeveloped, and the premaxilla can be floppy. The vermilion is thin in the lateral lip

手術後護理

傷口癒合和拆線

在修補雙側唇裂和初次鼻修復後，在患兒的唇部、兩邊人中柱皮膚、中線結節部份、內唇黏膜及或在鼻孔處有縫合線。

對於大多數患兒來說，傷口會在手術後一周左右癒合。在術後早期，如果護理不當，傷口可能會有負面影響。

每次餵食後，保持傷口清潔至關重要。家長在處理傷口前應先洗手。可用無菌棉籤和生理鹽水或冷開水去清潔傷口。可以用醫生處方外用藥膏如氯黴素軟膏，塗抹在傷口位置。如果在手術後貼了貼唇，只要唇貼乾燥、清潔和完好無損，直到拆線當天不需取下。

手術後，儘量避免反覆摩擦傷口。患兒需要配戴限制手肘活動的手套以避免搓揉傷口，但他們的手部不需要被拘束。要讓患兒感到舒適，避免他們過度哭鬧，以免影響上唇的傷口癒合。雖然不鼓勵患兒吸吮手指，但如果並無進行顎裂修補，應該不會有太大傷害。使用奶瓶餵食也要注意盡量減少對唇部造成壓力，如有需要可使用針筒或匙羹餵食。

患兒約在手術後 7 至 10 天入院拆線，但不同醫院安排各有不同。在大多數情況下，只需去除唇部皮膚上的縫線，而內唇和鼻子上的其餘縫線通常會隨著時間推移而被身體吸收。護士會指示父母在返到醫院拆線前禁食的時間， 因為患兒通常需要服用安眠藥（一般用水合氯醛）使他們安睡，以便拆線過程順利進行。

唇貼

唇貼是一種輔助手段去減輕傷口的張力，並起到保護性敷料的作用，術後貼唇貼的方法與手術前相約。如前所述，如果已經貼住唇貼，並在餵食後唇貼完好無損和乾爽，那麼在拆線前不需移除唇貼。不過，如果唇貼已髒，則應更換唇貼，貼上新唇貼前要清潔傷口。對於傷口良好癒合而言，唇貼並不是絕對必要的。因此，如唇貼貼得不好，家長無需焦慮不安，最好盡量避免過度觸到傷口。

餵食

為免在唇顎裂修補手術後餵食方法出現混亂，外科醫生在任何第一期修復手術（包括唇裂修復手術）後，建議家長使用針筒、滴管或匙羹去餵食。但實際上如只是唇裂修補而沒有進行顎裂修補的話，並沒有證明用奶瓶餵食有負面影響。餵食的原則是確保患兒得到充足的飽足而避免飢餓，因飢餓可導致患兒過度哭鬧。手術後，如患兒有鼻塞或嘴巴不能張大，使用針筒、滴管或匙羹餵食可能會更好。

segments. Presurgical nasoalveolar molding may not be able to help. Surgical technique of repair such cleft deformity may need to be modified and the primary repair result may be suboptimal. Secondary procedures are likely needed in such children, including adding dermal-fat and cartilage grafts.

Isolated bilateral cleft lip without cleft palate

Isolated bilateral cleft lip without cleft palate is not common. But if this occurs, the premaxilla may be solid, which makes presurgical nasoalveolar molding ineffective. If the premaxilla is in the way and affect lip repair, it may need to be set back first. The negative effect on midface growth may not be affected by the setback procedure given that the palate is intact.

Postoperative care

Wound healing and Stitch removal

After repair of the bilateral cleft lip and primary nose repair, suture lines are expected over the baby's lip, at lip skin at bilateral philtral column, midline tubercle region, inner lip mucosa, and possibly the nostrils.

For most babies, the wounds heal at around one week after the operation. During early postoperative period the wounds can be negatively affected if there is improper care.

It is essential to keep the wound clean after each feeding. Parents should wash their hands before taking care of the wound. Sterile cotton applicators can be used together with normal saline solution or cold boiled water. Topical ointment such as chloramphenicol ointment may be prescribed and to be applied to exposed area according to instruction. If the lip tapping was done after the operation, as long as it is dry, clean and intact, it may not be necessary to be taken off until the day of stitches removal.

It is important to avoid rubbing of the wounds repeatedly. The baby may need to wear special gloves to avoid manipulation of wounds. Their hands however do not need to be restrained. By making the baby comfortable, it can avoid excessive crying which itself can affect wound healing to the lip. Though suckling of finger is not encouraged, it should not be much harm if there is no palate repair done. It is the same for using bottle feed, although to minimize the risk of pressure to the lip, syringe or spoon feeding may sometimes be recommended.
The baby may be readmitted around day 7 -10 after the operation for stitches removal, depending on the practice of different centers. Most often,

鼻托

手術後，可以按照醫生的指示使用鼻托，可配戴數月以保持鼻型。可使用凡士林、溫水或嬰兒油等潤滑劑來幫助配戴。在餵食和晚間睡覺時可取出鼻托。鼻托每天至少要清洗兩次。如家長對患兒的呼吸有任何疑問，需馬上取出鼻托。鼻托是修復手術後維持鼻形的輔助工具。如在修復手術後鼻形良好，並不一定需要使用鼻托。

疤痕處理

拆線後，最重要的是在前 6-8 週內防止過度搓揉唇部。然而，鼓勵輕柔的按摩。有些人可能會考慮使用低過敏性膠帶，如醫生紙膠帶 (micropore) 或免縫膠帶 (steristrips)、矽膠啫喱或矽膠片作為輔助，減少疤痕的形成。

長期效結果

在最理想的裂唇修復情況下，嘴唇和鼻子應該仿似正常的唇和鼻，嘴唇輪廓光滑，疤痕不明顯，正常對稱的鼻部形狀、鼻樑均稱和輪廓分明的鼻尖。

然而，疤痕異常粗大或收縮很難完全預防。還有，幼童的疤痕最初是微細的差異，但隨著患者的成長可能會變得明顯。此外，與面部其他部分相比，鼻子、嘴唇和上顎骨的發育會比較遜色，從而出現第二期唇顎裂問題。

常見的第二期畸形包括以下幾種：
- 口哨狀唇形 / 上唇中央段不足
- 人中過闊
- 紅唇與人中交界處不對位
- 中面平坦
- 疤痕異常粗大

口哨狀唇形

對於上唇的中央段輕微不足而形成的口哨狀唇形，可以真皮脂肪移植去改善唇部輪廓。如上唇的中央段是頗為不足，可能需要使用下唇組織（阿貝氏瓣）分 2 個階段進行重建，使上及下唇的長度較為接近。

人中過闊

可能是由於第一次補唇手術時過度保留了前唇皮膚，或者由於突出的前齒骨持續拉扯導致上唇過闊或過長，這情況可以通過切除多餘的人中組織來糾正。

紅唇與人中交界處不對位

在第一期補唇手術後，如唇紅和人中交界處（白卷）有些不對位，可按差異的程度，透過不同的手術技巧進行糾正，包括切除和重新縫合、Z 形整形術、拆除修復和重新縫合白卷等。

only the stitches over the lip skin need to be removed and rest of the stitches over the inner lip and in the nose are often absorbable with time. The ward may instruct parents the timing of avoid feeding before coming back for stitches removal, because it is usually necessary for the baby to take sleeping medication, common chloral hydrate, to facilitate a smooth stitch removal procedure.

Tapping

Lip tapping is an adjunct for reducing the tension of the wound as well as serving as a protective dressing. The method of postoperative lip tapping may be similar to the preoperative ones. As mentioned above, if tapping was done and it is intact and dry after feeding, it may not be necessary to remove it until the day of stitches removal. However, if it is contaminated, it should be removed, and the wound should be cleaned before reapplying a new one. Lip tapping itself is not absolutely necessary for a good wound healing so there is no need to be anxious if somehow the lip tapping has not been ideal. It is best to avoid over-manipulation of the wounds.

Feeding

To avoid confusion of feeding method after lip and palate repair, some surgeons may prefer syringe, dropper or spoon feeding after any primary operation to the cleft deformity, though actually for lip repair only and without palate repair, there is no proven negative impact by bottle feeding. The principle of feeding is to achieve adequate feeding and avoid hunger of the baby which would also lead to excessive crying. In some cases the baby has nasal obstruction, or if the mouth of the baby is small after the operation, syringe, dropper, or spoon feeding may be better.

Nasal stent

After stitches removal, nasal stent may be used as instructed by the operating surgical team. It may be worn for several months after operation to maintain nasal shape. Lubricants such as Vaseline, warm water or baby oil may be used to facilitate insertion. It is taken out during feeding and at nighttime. It is to be washed as least twice daily. If there is any doubt concerning the babies breathing, do not hesitate to remove the stent. The stent is an adjunct to maintain the nasal shape after repair, and it may not always need the stent after a good nasal repair.

Scar management

After the stitches are removed, it is still essential to prevent over-manipulation of the lip for first 6-8 weeks. Gentle massage however is encouraged. Some may consider use hypoallergenic taping such as micropore or steristrips, silicone gel or sheet as adjunct for minimizing scarring.

中面平坦

唇顎裂患者的上顎骨發育不良是可以預期的，牙列和咬合亦可能會受到影響，從而可能需要進行矯齒或正顎手術。在下顎骨矯正手術完成後，可考慮進行唇鼻整形手術。

疤痕異常肥大

任何手術後都會產生疤痕。最理想的情況是疤痕不顯眼。疤痕的發展範圍很廣，從幼細成熟疤痕到粗厚性疤痕，以至蟹足腫疤痕。

手術後幾個月要避免日曬，疤痕按摩有助防止過度色素沉殿和減少疤痕的厚度。使用矽膠啫喱或矽膠片可能有助減少副作用。對於較為嚴重的疤痕可注射類固醇或經醫生評估使用其他疤痕處方法。

Long term Outcome

Ideally, the reconstructed lip and nose should mimic normal ones, with smooth lip contour, inconspicuous scar, symmetrical nostrils of normal shape, presence of adequate columella, and well-defined nasal tip.

However, scar hypertrophy or contraction is difficult to totally prevent. Also, growth of the patient may make an initially minimal discrepancy in very young kid to become conspicuous when they have grown up. Furthermore, certain elements of the nose, lip, and maxillary skeleton are expected to be less well developed compare to rest of the face.

Common long-term deformity includes the followings

- Whistle deformity/Central segment deficiency
- Widen philtrum
- Mismatch at skin-vermillion junction
- Flat midface
- Hypertrophic Scar

Whistle deformity

For minor central segment deficiency, dermal fat graft may be useful to camouflage the lip contour. If there is significant deficiency, It may be necessary to reconstruct in 2 stages using lower lip tissue (Abbe's flap).

Widen Philtrum

It may be due to over-preservation of prolabium skin at the primary surgery, or due to persistent stretching by a protruding premaxilla. It may be corrected by excision of excess philtral tissue.

Mismatch at lip-vermillion junction

A small mismatch at the lip-vermillion junction (white roll) may be noticeable from long distance. Depends on the level of mismatch, it may be corrected by different methods include excision and resuturing, z-plasty, to taking down the repair and reapproximation of the white roll.

Flat midface

Undergrowth of maxilla is anticipated. Dentition and occlusion may be affected such that orthodontics and orthognathic surgery may be necessary. Soft tissue surgery such as lip-nose revision is to be consider after the underlying bone foundation is settled

Hypertrophic scar

Scar always occur after any surgery, and the best scenario is it remains inconspicuous. There is a wide spectrum of scar condition ranging from fine mature scar to hypertrophic scar and keloid formation. Avoiding sun exposure and massage of scar several months after operation may be helpful to prevent hyperpigmentation and decrease the thickness of the scar. Silicone gel or sheet may be useful and has minimal side effect. For more severe scarring, intralesional steroid injection or other modalities of scar management would depends on medical assessment.

全裂及不全裂顎的修補手術
Complete and Incomplete Cleft Palate Repair

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儘管顎裂與唇裂相比是一種較為隱蔽的異常，並且外觀上對父母來說並不是令人震驚，然而顎裂對多種身體功能帶來影響，包括餵食和說話。適當和及時的手術修復對上顎的長期功能起重要決定性作用。本文將重點介紹顎裂的處理和修復手術。

Although cleft palate is a more conceal deformity and not as cosmetically alarming to parents when compare to cleft lip, it has several functional implications including feeding and speech. Proper and timely surgical repair will determine the long term function of the palate. This article will focus on the management and surgery for cleft palate.

在胚胎學上顎骨的發育

在胚胎學上，從門齒孔而發展上顎可分為初生顎和次生顎。初生顎在妊娠大約第五週，從內側鼻突發育，形成上唇和上顎，包括中門齒。它將合併到側鼻突發育的鼻子和嘴唇。次生顎在妊娠第 6 週開始發育，到第 12 週完全形成。最早於上頜突開始，分離的顎板向內側遷移並從門齒孔前後合併到吊鐘，形成軟和硬的上顎。

唇顎裂的成因有幾種理論可以解釋。目前，公認的唇顎裂成因是神經外胚層細胞遷移失敗所致。神經外胚層細胞負責在外胚層裂解後合併面部過程。

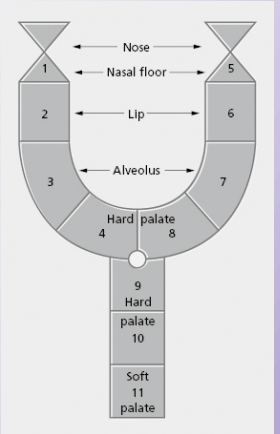
Embryology of palatal development

The palate is embryologically divided into primary and secondary palate by the incisive foramen. The primary palate develops around the fifth week of gestation from the medial nasal process, forming the upper lip and the upper jaw including the central incisors, which will merge to the lateral nasal processes, the nose and the lip. The secondary palate begins developing during the 6th week of gestation, and is completely formed by the 12th week. It originates from the maxillary process in which the separated palate lamina migrates medially and merges anterior-posteriorly from the incisive foramen to the uvula, forming the hard and soft palates.

There are several theories to explain the origin of cleft lip and palate. It is currently accepted that cleft lip and palate is due to a failure in the migration of neuroectodermal cells, which is responsible for merging the facial processes after ectodermal lysis.

顎裂的分類

克納漢和斯塔克醫生的分類方法 (Kernahan and Stark's classification)，系統在其中心內被採納。它是有效表達唇裂和顎裂情況的方法。克納漢醫生的 Y 型圖 (Kernahan "striped-Y") 的設計主要是為了簡化記錄保存。但它也使分類成為一種視覺作用，而不是一種抽象的認知。克納漢醫生 Y 分型圖 (Kernahan "striped-Y") 以及埃爾薩希醫生 (1973) 和米拉德醫生 (1976) (Elsahy and Millard) 的後期修改，有助於確保克納漢和斯塔克醫生的分類法 (Kernahan and Stark's classification)，作為日常臨床使用的主要系統。



Classification of cleft palate

Kernahan and Stark's classification is the system used in author's center. It acts as an effective way of communication for the extent of the cleft lip and palate. The Kernahan "striped-Y" diagram was designed principally to simplify record keeping, but it also made classification a visual process rather than an abstract, cognitive exercise. Kernahan's striped-Y and the subsequent modifications by Elsahy (1973) and Millard (1976) served to secure the position of the Kernahan and Stark classification as the predominant system used in daily clinical practice.

對於不全顎裂，我們所指的是不同程度的顎裂患者而不涉及齒槽的，如吊鐘裂、軟顎裂和硬顎裂。在單側顎裂情況下，上顎的一側附著在犁骨上。在雙側顎裂情況下，上顎兩側均不附著於中央犁骨。完全次生顎裂患者的縫隙裂至前門齒孔。對於單側完全唇顎裂，只有一側牙槽裂。如果是雙側完全唇顎裂，雙側牙槽都會有裂隙，而初生硬顎將與上顎的其餘部分完全分離。

功能性異常及術前管理

顎裂患兒由於無法有效地吸吮和鼻腔倒流而導致餵食困難，可使用長奶嘴和軟身瓶以方便餵食。在公立醫院，嬰兒餵哺隊包括哺乳顧問和職業治療師會在嬰兒出生後儘快協助母親餵哺患兒。

因患兒的耳咽管功能不佳，使他們較容易出現中耳積液，所以在患兒出院時會轉介耳鼻喉科醫生跟進。長期中耳積液不管有沒有積液性中耳炎都會影響孩子的聽力發育。在上顎修復時，應同時植入中耳導管以流放積液。

上顎肌肉的另一個重要功能是在說話時收縮而達至顎咽完全閉合。遲遲未修復的顎裂會導致患兒說話有鼻音，這個語言問題是在患者成年後很難糾正的。當孩子開始說句子並懂得配合訓練時，他們會被轉介給言語治療師進行訓練。經過一段時間的訓練後，如仍無法達到令人滿意的語言能力，患者將被轉介去作進一步顎咽閉合不全的檢查，看看是否需要手術來矯正。

牙槽裂可能導致牙齒問題，進而需要接受治療。患童將被轉介給牙科外科醫生，以評估牙床植骨手術的需要，以免在換牙期時影響犬齒的生長。牙科外科醫生稍後將評估患者是否需要牙齒矯正療程和正顎手術，以糾正牙齒咬合和面部輪廓。但對於原本有顎咽閉合不全的患者來說，在正顎手術時上顎被前移後，他們的顎咽閉合不全問題可能會更為嚴重。因此，包括語言治療師、牙科外科醫生、整形外科醫生等的跨專科綜合治療是唇顎裂治療的標準模式。

For incomplete cleft, we are referring to those patient with different degree of cleft palate not involving the alveolus i.e. bifid uvula, cleft soft plate and cleft hard palate. In unilateral case, one side of the palate is attached to the vomer. In bilateral case, both sides are not attached to the central vomer. Those patients with complete cleft secondary palate will have their cleft reaching the incisive foramen anteriorly. For unilateral complete cleft lip and palate, one side of the alveolus is involved. In case of bilateral complete cleft, alveolus will be involved bilaterally and the primary hard palate will be completely detached from the rest of the palate.

Functional abnormality and pre-operative management

Feeding difficulties due to inability to suck effectively and nasal regurgitation. Special feeding bottle with long nipple and soft bottle can be used to facilitate feeding. Infant feeding team including lactating consultant and occupational therapist will be attending the patient and mother to facilitate feeding soon after the baby is delivered.

ENT surgeon should be referred upon discharge because patient will be prone to middle ear effusion due to the malfunctioning of Eustachian tube. The prolong middle ear effusion with or without repeated otitis media will affect the hearing development of the child. Definitive treatment including myringotomy with insertion of grommet should be performed during the repair of palate.

Another important function of the palate is to achieve a complete velopharyngeal closure when the muscle contract during speech. Persistent unrepaired cleft palate will result in nasal speech, which is very difficult to correct in adulthood. Children will be referred to speech therapist for training when they start speaking sentences and can cooperate with training. Those who cannot achieve satisfactory speech after a period of training will be referred back for further investigation for velopharyngeal incompetence, which warrant surgical correction.

The involvement of cleft alveolus may result in dental abnormality requiring treatment. Children will be referred to dental surgeon to assess the need of alveolar bone grafting to facilitate the crowning of the affected canine at the time of mixed dentition. The patient will be assessed later for the need of orthodontic treatment and orthognathic surgery to correct the occlusion and facial profile. However, for those patient with velopharyngeal incompetence, orthognathic surgery may further worsen the condition during the process of moving the maxilla forward. Therefore a multidisciplinary approach involving speech therapist, dental surgeon and plastic surgeon is the standard of care in managing patient with cleft.

Surgical techniques of palatoplasty

裂顎修復手術技巧

手術時間取決於 10 的規則，例如：患兒要有 10 週歲大，血紅蛋白有10 公克和體重達至 10 磅。一般來說，整形外科醫生通常在患兒 9-12 個月大，最好是在 2 歲之前進行顎裂修復手術。但是，針對綜合症群患者的手術可能要根據他們的整體情況和語言發展進度而作出特別安排。

顎裂修復手術會在全身麻醉下進行。氣道插管會置於中線下方並以膠布固定位置。患兒會被放在手術台的最末端位置，讓頸部伸展和以肩部支撐以方便手術進行。外科醫生可以坐在患兒的頭端或右側位置。使用丁曼口腔張開器使口腔黏膜和舌頭退後，以露出手術位置，同時確保氣管導管供應充足氧氣。用凡士林紗布保護口角，以防止由於嘴唇過度拉開而導致撕裂。進行喉嚨填塞以防止麻醉氣體洩漏和血液吸入。當位置準備好，切口將被注射利多卡因 + 腎上腺素，以增強在手術過程中的止血功效和控制術後疼痛。

雙瓣顎裂修復技術

Janusz Bardach 於 1967 年首次描述了雙瓣顎裂修復技術。目的是達到一次性兩層式的修復整個硬顎和軟顎。

手術的第一步包括設計顎裂邊緣處的切口，以形成黏膜骨膜瓣。上顎裂隙越闊，留在鼻後的黏膜骨膜邊緣會越大。位於上顎邊緣的這條黏膜骨膜帶將向下轉移以閉合鼻底。切口以編號 12 刀片從軟顎中間邊緣開始。切口將向前延伸以連接在上顎和牙槽黏膜之間的硬顎切口，然後，可以使用細尖骨膜剝離器來分割口腔側的黏膜骨膜瓣，分割直到硬顎後邊緣，以達至較大的顎孔。使用黃氏裂顎解剖器，將神經血管束從周圍肌肉中分離。用銳利的解剖器，使軟顎肌肉完全從硬顎後邊緣和口腔及鼻黏膜中完全分離出來，使鼻黏膜骨膜被大幅度往下打開。用 4/O 號縫線從上牙槽開始把鼻底和顎裂縫合。把缺裂的提顎帆肌和張顎帆肌以雙排式縫線接駁。上顎肌肉的閉合將從吊鐘尖端向前進行。

把肌肉適當地對位後，口腔黏膜的縫合就變得很容易。口腔黏膜骨膜瓣應縫合到硬顎上方的鼻層。這可防止層與層之間留下死腔。將兩個黏膜骨膜瓣的前部縫合到原生硬顎，以防止其下垂。我會在吊鐘後面放

The timing of operation is determined by the rule of 10 i.e. 10 weeks of age, 10 grams of haemoglobin and 10 pounds. In general, we usually repair the palate at around 9-12 months old, preferably before 2 years of age. However, operation for syndromal patient may be tailor-made according to their general condition and speech development.

The patient will be operated under general anaesthesia. Endotracheal tube should be placed inferiorly in the midline and tapped to secure the position. Patient should be put at the very end of the operating table with neck hyperextended with shoulder support to facilitate operation. Operating surgeon can be seated at the head end or right side of the patient. Dingman retractor will be used to retract the buccal mucosa and tongue, to expose the operating site while making sure the ventilation via the endotracheal tube is adequate. Oral commissure will be protected by Vaseline gauze to prevent laceration due to over-stretching of the lip. Throat packing was performed to prevent leaking of anaesthetic gas and aspiration of blood. Once the position is ready, the incision will be injected with lignocaine + adrenaline to enhance haemostasis during operation and control post op pain.

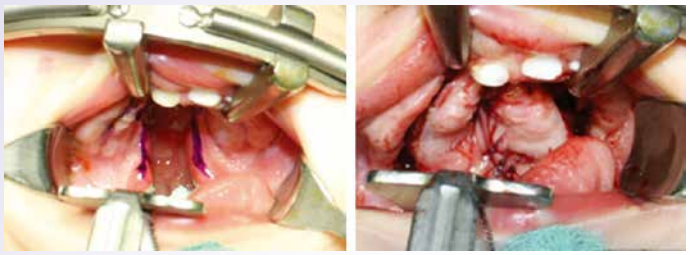
Two-flap palatoplasty

The two-flap palatoplasty technique, which Janusz Bardach was described for the first time in 1967. The aim is to obtain a two-layer closure of the entire cleft hard and soft palate in single stage.

The first step of the operation involves designing the incisions at the cleft margins for creation of the mucoperiosteal flaps. The wider the palatal cleft, the larger the margin of mucoperiosteum that must be left at the nasal side of the palatal cleft. This strip of mucoperiosteum at the palatal edge will be turned downward for closure of the nasal layer. The incision starts along the medial margin of the soft palate with blade number 12. The incision will be extended anteriorly to connect with the hard palate incision between the palate and alveolar mucosa. Then a fine tip periosteal elevator can be used to undermine the mucoperiosteal flaps on the oral side. Dissection continue until the posterior edge of the hard palate and the greater palatine foramen are identified. Using a Wang cleft palate dissector, the neurovascular bundle is bluntly dissected from the surrounding muscle. The muscles of the soft palate are then completely released from their attachments at the posterior edge of the hard palate and from the oral and nasal mucosa with sharp dissection. The nasal mucoperiosteum is widely undermined. Closure of the nasal layer of the palatal cleft begins at the alveolar ridge with 4/O Vicryl. The levator and tensor muscles of both sides are sutured together in double breast manner. Closure of the muscles will proceed from the tip of the uvula forward. After the proper apposition of muscle, closure

1 或 2 針抗張力縫合線，以保護患兒在哭鬧時吊鐘傷口裂開。

黏膜骨膜瓣兩旁露出的骨骼應用可完全吸收的止血棉填充，以加強止血功效和防止骨骼枯乾。



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雙層反向 Z 形顎裂修補術

雙層反向 Z 形顎裂修補術的基本原理是採用兩層反向 Z 型成形術來對顎肌肉進行移位。對於慣用右手的外科醫生，後部口腔黏膜皮瓣位於左側。鼻腔的 Z 型成形術有如口腔層的鏡像反映。兩側口腔 Z 型成形術的側肌完成呈小鉤狀。兩個肌黏膜瓣向後轉置，另外兩個黏膜瓣向前轉置。口腔和上顎黏膜將用 4/O 號針線縫合。以這種方式閉合的上顎將在前後方向上拉長，但代價是令到兩邊黏膜在縫合時會特別繃緊。至於延長程度則取決於軟顎側肌與中央中線肌肉的角度 - 通常在 45°-60° 之間 (達到 50-75% 的伸長率)。

雙層反向 Z 形顎裂修補術適用於軟顎裂、隱性顎裂和由於軟顎短或功能不佳而導致的顎咽閉合不全症狀。

手術後，應清除口部咽喉的血液。確認止血後，應取出喉部紗布。丁曼口腔張開器會小心取出，在過程中特別注意不要取出氣道插管。

術後最初的 24 小時內，應密切監測患者的術後出血情況。術後最初幾天唾液帶有血是很常見。待患者完全清醒後，可開始以口腔餵食。最初患兒可以進食流質，然後在兩週內逐步過渡至軟質飲食。為了防止新修補的硬顎受損，我們會建議家長使用針筒或杯子餵食。術後要經常喝水保持口腔衛生。使用常規必理痛糖漿，可以控制傷口疼痛。

of the oral mucosa will usually be easy. The oral mucoperiosteal flap should be sutured to the nasal layer over the hard palate. This prevents leaving a dead space between the layers. The anterior part of the two mucoperiosteal flap will be sutured to the primary hard palate to prevent it from hanging down. I like to put 1 or 2 tension suture behind the uvula to protect the wound gapping when the baby cry.

The exposed bone lateral to the mucoperiosteal flap will be packed with Surgicel to promote haemostasis and prevent the dessication of the bone.

Furlow palatoplasty

The basic principles for the Furlow's palatoplasty were transposition of the palatal muscles with double opposing z-plasty. The posteriorly based oral myomucosal flap was on the left side for a right handed surgeon. The nasal Z-plasty was made as the mirror image of the oral layer. The lateral limbs of the oral Z-plasty ended over the hamuli. Two myomucosal flaps are rotated posteriorly and the two mucosal flaps are transposed anteriorly. Oral and palatal mucosa will be closed with 4/O Vicryl. Palate closed in this manner will be lengthened in anteroposterior direction at the expense of increased tension in closure of the lateral mucosa. Degree of lengthening is dependent on the angle of the soft palate lateral limbs from the central midline limb - usually between 45°-60° (50-75% elongation achieved).

The indications for Furlow palatoplasty are cleft soft palate, submucosal cleft and velopharyngeal incompetence due to short or poorly functioning soft palate.

After the operation, blood should be cleared from the oropharynx. Throat pack should be removed after confirming the haemostasis. Dingman retractor will be removed with care, paying particular attention not to remove the endotracheal tube during the procedure.

Patient should be kept in close monitoring for post-operative bleeding for the first 24 hours. It is very common to have blood stained saliva for the first few days. Oral feeding can be started when patient is fully awake. Initially patient can try fluid diet and then step up to soft diet in two-week time. In order to prevent damage to the newly repaired hard palate, we will recommend parents to feed with syringe or cup. Oral hygiene should be kept with frequent drinking of water. Pain control is usually adequate with regular syrup panadol.

Velopharyngeal Incompetence

顎咽閉合不全

患童在 5 歲後或當他們能夠配合時，可以接受顎咽閉合不全的評估。患者通常會有說話時鼻音重的情況，他們會首先由語言治療師進行評估及治療，以糾正其他同時出現的問題，如：發音問題。如問題未有改善，患童需接受相關檢查，包括鼻咽喉內窺鏡和視頻透視吞嚥檢查，以評估患童的顎咽閉合不全原因和嚴重程度。如結構性不足的患童可能需要手術矯正。雙層反向 Z 形手術、咽喉瓣手術、括約肌咽成形術或注射式咽成形術可用於改善顎咽閉合。

皮爾羅賓症候群

皮爾羅賓症候群是一種罕見的疾病，臨床表現為小下巴、舌下垂和裂顎三種症狀合成口腔頜面的異常。拉納隆格和梅納德 (Lannelongue and Menard) 於 1891 年首次描述了皮爾羅賓症候群。在 1926 年，皮爾羅賓 (Pierre Robin) 發表了一個嬰兒患有完全皮爾羅賓症候群的案例。直到 1974 年，這三種症狀被稱為皮爾羅賓氏序列畸形。醫學用語「症候群」現今用於由一個畸形所引發的一系列問題。皮爾羅賓症候群的確切病因尚未清楚。這種先天性缺陷的患病率約為每 8500 名新生嬰兒中會有 1 名。男女比例為 1:1，X 染色體遺傳除外。它可以獨立出現，也可以是一些綜合症狀的一部分。其中最常見的綜合症狀包括斯蒂克勒症候群 (Stickler syndrome)、顎 - 心 - 臉症狀群 (velocardiofacial syndrome)、腦 - 肋 - 骨與下頜骨症候群 (cerebrocostomandibular syndrome)、愛德華氏症 (trisomy 18) 和胎兒酒精症候群 (fetal alcohol syndrome)。

力學理論是皮爾羅賓氏序列畸形最普遍接受的病理生理學。最初期，專家發現有些胎兒全在妊娠的第 7 周和第 11 週之間下顎發育不良。這使舌頭在口腔中保持高位置，阻礙上顎的合在一起而導致顎裂問題。這個理論解釋了經典的倒 U 形顎裂而沒有陪伴唇裂。在病因學而言，羊水過少可能是原因之一，因為缺乏羊水會導致下巴變形，以致舌頭受壓塞於顎架之間，影響上顎的閉合。

小下巴和舌下垂同時出現可能導致新生嬰兒嚴重的呼吸和餵食困難。可讓患兒俯伏式躺著，以保持呼吸道暢通。如俯伏式躺著仍不足夠，可考慮其他選項如口腔導氣管、喉罩、鼻咽支架或短期插管。

Patient will be assessed for velopharyngeal incompetence after age 5 or when they can cooperate. Patient usually presented with hypernasal speech and they will be first assessed by speech therapist to correct any other coexisting condition such as articulation problem. Investigations including nasopharyngolaryngoscopy and video fluoroscopic swallow study can be used to assess the cause and severity of the condition. For those patients with anatomical deficiency, surgical correction may be needed. Surgeries include Furlow palatoplasty, posterior pharyngeal flap, sphincter pharyngoplasty or injectable pharyngoplasty can be offered to improve the velopharyngeal closure.

Pierre Robin Sequence

Pierre Robin sequence is a rare disease presenting with the clinical orofacial abnormality triad of micrognathia, glossoptosis, and cleft palate. Lannelongue and Menard first described Pierre Robin syndrome in 1891. In 1926, Pierre Robin published the case of an infant with the complete syndrome. Until 1974, the triad was known as Pierre Robin syndrome. The term sequence was now used to include a series of anomalies caused by a cascade of events initiated by a single malformation. The exact etiology of Pierre Robin Sequence is unknown. This heterogeneous birth defect has a prevalence of approximately 1 per 8500 live births. The male-to-female ratio is 1:1, except in the X-linked form. It can exist independently or as part of a number of other syndromes. The most common of these are Stickler syndrome, velocardiofacial syndrome, cerebrocostomandibular syndrome, trisomy 18, and fetal alcohol syndrome.

The mechanical theory is the most commonly accepted pathophysiology of Pierre Robin Sequence. The initial event, mandibular hypoplasia, occurs between the 7th and 11th week of gestation. This keeps the tongue high in the oral cavity, causing a cleft in the palate by preventing the closure of the palatal shelves. This theory explains the classic inverted U-shaped cleft and the absence of an associated cleft lip. Oligohydramnios could play a role in the etiology since the lack of amniotic fluid could cause deformation of the chin and subsequent impaction of the tongue between the palatal shelves.

The combination of micrognathia and glossoptosis may cause severe respiratory and feeding difficulty in the newborn. Baby can be nursed in prone position to protect the airway. Oral airway placement, laryngeal mask, nasopharyngeal stenting, and short-term intubation are other options in case prone positioning is inadequate.

Feeding difficulties can be alleviated by upright feeding techniques, modification of the nipple for bottle feeding, temporary use of feeding tube. In

餵哺困難可透過坐直式餵奶姿勢、奶嘴調整或暫時使用胃喉餵食來緩解。在嚴重的情況下，可能需要放置胃造口以便餵食。帶有前會厭棒板的顎板是一種有用的輔助工具，有助把舌根向前拉。這種方法可以擴大氣道，並促進吞嚥。

鑑於顎裂修復後可能出現呼吸問題，9-12 個月的常規顎裂修補時間可能不適用於這些患兒。修補時間應與兒科醫生商討，以配合個別患兒的情況。有時顎裂修復可能需要推遲到 1 歲以後，但最好在 2 歲之前。對於那些有綜合症狀的患兒，可以根據患者的一般情況和語言發展進一步推遲顎裂的修補。

由於顎裂縫隙較為寬闊，皮爾羅賓症候群患兒的顎裂修復手術頗為困難。小下巴和舌下垂會使患兒上顎難以打開，使情況進一步複雜化。當使用雙瓣顎修復技術時，最繃緊的部分通常是硬顎和軟硬顎交界處的鼻黏膜。在這種情況下，可以犁骨瓣促進鼻黏膜的閉合。兩邊式犁骨瓣可用於閉合硬顎前端部份。單或雙的後端犁骨瓣可以轉動以關閉軟硬顎交接處的鼻黏膜。口腔黏膜骨膜瓣的閉合通常在椎根骨架化和解除切口向外側延伸到椎根後部延伸後完成。雖然口腔和鼻腔黏膜之間的死腔可能無法在術後即刻被消除，但通常會在術後順利癒合。分層閉合的目的是防止漏管形成。然而，這可能不適用於每一個寬闊顎裂病例。在極端情況下如果有需要，可以使用口腔瓣來促進顎裂閉合。

結論

顎裂修補是一項改變生命的手術，如果手術進行妥當，可使患者去標籤化，讓他們更好地融入社會。

severe case, gastrostomy tube placement may need to facilitate feeding. Palatal plates with pre-epiglottic baton plate is a useful adjunct to help to pull the base of the tongue forward. This maneuver can widen the airway and facilitates swallowing.

In view of the possibility of airway problem after the closure of cleft palate, routine repair of the cleft palate in 9-12 months may not be applicable to this group of patients. Timing of repair should be discussed with paediatrician to suit individual patient's need. Sometime it may need to be delayed beyond 1-year-old, but preferably before 2 years of age. For those syndromal patient, repair of cleft palate can be delayed further according to the patient's general condition and speech development.

Surgery for cleft palate in patient with Pierre Robin Sequence will be difficult because of the wide cleft palate. Micrognathia and glossoptosis will make the exposure difficult, which further complicate the situation. When performing closure with two flap palatoplasty, the tightest part is usually the nasal mucosa over the hard palate and the hard soft palate junction. In those case vomer flap can be used to facilitate the closure of nasal mucosa. Two sided vomer flap can be used to close the anterior part of the hard palate. Single or double posterior based vomer flap can be turned to close the nasal layer of the hard soft palate junction. The closure of oral mucoperiosteal flap is usually achievable after skeletonization of the pedicle and posterior extension of the releasing cut laterally beyond the pedicle. Although the dead space between the oral and nasal mucosa may not be able to be obliterate during the immediate post-operative period, it usually healed uneventfully afterwards. The aim for layered closure is to prevent fistula formation. However, it may not be possible in every single case of wide cleft palate. In extreme case, buccal flap can be used to augment the closure if needed.

Conclusion

Cleft palate repair is a life changing surgery, if performed properly, can remove the stigmatization and allow patient to integrate better to the society.

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小耳症修補手術
Microtia: Where Are We Now?

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小耳症 (microtia) 泛指所有外耳畸型問題，包括最輕微的外耳形狀和大小異常以致完全缺乏外耳 (無耳症)。小耳症可能單獨發生，亦可能是基因性的症群。小耳症是胚胎期形成外耳時，外胚層的胚胎細胞產生病變所造成的。目前醫學上發生病變的原因不明，父母不必為此而感到內疚。小耳症發生在男性的比率較女性為高，約佔總數的 60-70%。大部分為單側小耳症 (比例為 80-90%)，其餘為雙側性小耳症。亞洲地區之發生率約為 5000 分之一。由於聽力和外觀均受影響，兒童早期發展時應尋求耳鼻喉科醫生做聽力診斷與跟進，並且由整型外科醫生就外耳的重建手術進行評估、分析治療方案。

重建外耳確對患者來說非常重要，因為不單可以改善外觀，亦可方便配戴眼鏡和口罩，更重要是幫助患者建立自信，減低社會心理影響。自 1950 年代起，不斷有關專家提出手術計劃建議。在這許多年的發展中，不斷改良手術方法，以減低病人需經歷的手術次數，亦透過 3D 掃描和打印技術的輔助，以爭取更好的耳廓重建效果。

外耳重建的目的在於提供外觀的完整性。重建方式主要分為自體組織 (肋骨軟骨) 移植重建、 聚乙烯人工軟骨 (Medpor) 或配戴耳朵模型。

1) 自體軟骨移植

以自體軟骨移植來重建耳朵是目前最普遍及最安全的選擇。現今主流是由日本的永田悟醫生 Dr.Satoru Nagata 發明的二階段外耳自體組織移植重建手術，稱之為永田悟手術。第一階段先取出肋骨軟骨雕出耳型，此時就擁有一個平面耳朵，半年後進行第二階段把耳朵掀起，再將肋骨墊於耳朵後使之豎立。手術最好等患者到 10 歲左右進行，才有足夠之軟骨來雕刻外耳。隨著技術發展，個別病人在第一階段手術後，新耳朵已能夠豎立，足夠戴眼鏡或口罩，而無需再進行第二階段手術。然而由於牽涉取肋骨，儘管不影響功能，術後仍會有疼痛及留下疤痕。

2) 人工骨 (Medpor)

採用人工骨 (Medpor) 用作外耳重建亦是新的方式。Medpor 的結構是多孔性的，目的是讓人體的組織

Microtia refers a wide spectrum of malformations of the external ear, ranging from minor irregularities in the contour and size of the ear to a complete absence of the ear, known as anotia. Microtia can occur in isolation, or as part of a genetic syndrome. The aetiology remains unclear. The incidence in Asian population is about one in 5000. Microtia is more common in males which comprises about 60-70% of all the affected. The majority are unilateral (80-90%) while the remaining are bilateral. In terms of hearing impairment, in those patients with unilateral microtia, the majority of patients have normal hearing in the unaffected ear and hence normal conversation are not affected. For those with bilateral involvement, early ENT consultation and audiologist assessment are crucial for speech and intellectual development.

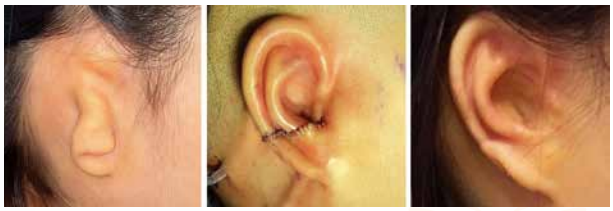
Reconstruction of the external ear plays an essential role not only in rebuilding the ear for appearance, for wearing spectacles or masks, but more importantly, in building up one's confidence and eliminating the psychosocial stigma. Various strategies have been proposed since 1950s. Modifications were made throughout the years to decrease the stages that patient has to go through, while efforts were made in achieving the best possible reconstruction outcomes with adjunct of 3D scanning and printing technology.

Current available options of reconstruction are:

1) Reconstruction using costal cartilage

A two-staged operation was proposed by Professor S. Nagata in 1993. First stage can be done when one reaches 10 years old with chest wall circumference more than 60cm. The costal cartilages are fabricated into an ear shape and then placed underneath the ear skin. After at least six months, second stage is performed. The ear is elevated and projected with a cartilage block and covered by a well-vascularized fascial flap and skin graft. This is a safe operation with a longstanding history over decades, as the newly fabricated ear actually comes from the patient's body with low risk of infection or exposure. Chest wall deformity is minimal and would not affect respiratory function or daily activities after recovery. With modification of techniques, in selected patients with favourable

自體軟骨移植重建
Reconstruction using costal cartilages



手術前 (左), 即時手術後 (中) 和手術後半年 (右)。
Lobular type microtia before (left) and immediate after operation (right).

或血管能夠生長進這些微小的孔洞，使植入的 Medpor 不易移位，同時也降低其感染與排斥的機率。由於不需肋骨軟骨，手術可提早至五至六歲進行。 手術時必需要從耳上方的頭皮下取出一塊筋膜和植皮來覆蓋著人工骨，才能確保人工骨有血管及組織保護，不致外露。可是，萬一這個覆蓋不健全，可想而知，人工骨便有外露的風險，嚴重者甚至會有細菌感染發炎的情況，甚至乎最後需要取出已放置的人工骨才能控制炎症。

3) 外掛式義耳

可採用外掛式義耳黏附在耳朵位置，或使用嵌入鋼模式或磁性義耳。這種人工義耳會隨著時間而磨損、變舊，亦可能需要醫生定期跟進和調整。同時長久使用黏貼性義耳可能會導致皮膚敏感和發炎。嵌入鋼模式義耳並且需要小心護理，以免引致併發症。

總結

不同類別的耳廓選項有其優點，亦有其缺點，最重要是經過詳細手術前評估及諮詢病人和家屬後，才決定那一個選項最適合個別病人，同時亦視乎負責手術的醫生的經驗。任何重建手術，第一次手術是非常重要的，因為要確保健康的皮膚組織得以善用。否則，若日後皮膚變成了疤痕，重建整型的效果便會稍遜。故此，手術前的諮詢和策劃、外科醫生的經驗尤其重要。希望各小耳症的大小朋友也能早日找到合適的方案，讓小耳朵重新站起來！

最後，在這新時代，小耳重建仍然是在不斷發展中。近年，不斷有專家在研究培植軟骨細胞或幹細胞，再以 3D 生物科技打印耳廓，希望在未來能夠移植到病人身上，這樣就可以做出更加對稱的、形狀接近正常的、靈活而有富彈性的、安全和穩定的、終生可使用的耳廓，造福所有小耳症患者。

手術前 (左) 和手術後一年 (右)。
Lobular type microtia before (left) and one year after operation (right).

conditions, they can indeed put on spectacles or masks after the first stage and hence sparing the need for second stage operation.

2) Porous implant reconstruction using Medpor

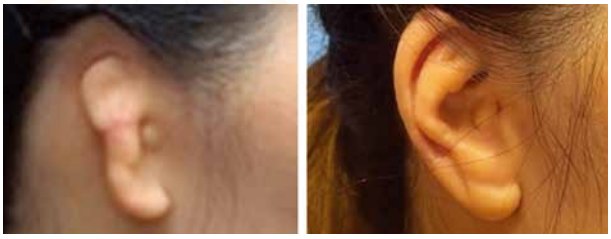
Medpor is a high density polyporous polyethylene material that allows tissue and vascular ingrowth. The entire implant requires complete soft tissue coverage to ensure success, i.e. a thin fascial raised from underneath the scalp. Since there is no need for cartilage harvest, operation can be advanced to age 5 to 6 depending on patient's readiness and surgeon's experience. Chest wall scars are eliminated, nevertheless, complications would include wound problems, infection, implant exposure or fracture, and in the worst situation, removal of implant may be necessary to control the damage.

3) Prosthetic ear

Prosthetic ear can be adhered to skin using adhesives, or in the form of osseointegrated implants or magnets. However, there would be wear and tear with time and hence, one would require regular follow-up and adjustment if needed. Furthermore, dermatitis may result due to use of adhesives. The osseointegration abutment also requires proper care to avoid complications.

In summary, different options of reconstruction have their own advantages and disadvantages. It is important to have a detailed preoperative assessment and thorough counselling with patients and their family, in order to decide on the most suitable option for individual patients. Satisfactory reconstruction outcomes is achievable on experienced hands.

Last but not least, tissue engineering, the blending of biology and engineering, is a developing field in the new era of reconstruction. Research is now ongoing to evaluate the application of cartilage cells or stem cells on 3D-printed biocompatible ear scaffold which can, hopefully, be implanted under skin in the near future, with the aim to create an ear which is symmetrical to the normal ear, flexible and elastic in consistency, and remain safe and stable lifelong.



兔唇裂顎患者的言語治療問題及處理
Speech Therapy for Children with Cleft Lip and/or Palate

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語音特徵

兔唇裂顎兒童有言語問題是十分常見。兔唇顎裂對他們的言語發展有負面影響，部份患兒可能有〝顎裂語音〞，伴有非典型輔音產生、異常的鼻腔共振和氣流。語言問題包括咽塞音、咽擦音、鼻發出的壓力輔音和舌顎塞音。此外，經常出現過度的鼻腔振動使鼻音過重，兔唇裂顎典型語音特徵如下：

1. 言語發展延遲（壓力和非壓力輔音）
2. 語音清晰度降低
3. 遺漏和替代音比扭曲音更頻繁
4. 喉音和咽音的替代或其他〝上顎後方〞產生的語音（例如：牙槽至軟顎或聲門等）
5. 鼻音替代

言語障礙的治療和管理

兔唇顎裂兒童言語障礙的治療方法，包括外科手術和言語治療。外科手術針對的是結構異常和顎咽閉合不良而導致語音偏差。手術修復後，兔唇裂顎兒童需要進行言語治療矯正可能已出現代償性的語音錯誤。下面列出常用的外科手術和言語治療的技術：

A. 外科手術

1. 裂唇修補手術（通常在出生後的 6 個月內進行）
2. 裂顎修補手術（通常在正常發育 12 個月大的兒童進行）
3. 隱性顎裂修補手術
4. 矯正非裂顎咽喉疾病或矯正術後顎咽閉合不良的手術（例如：咽瓣膜手術、括約肌咽成形手術、顎延長手術、咽壁內部擴張手術）
5. 扁桃體切除手術和 / 或腺樣體切除手術（可能在進行咽瓣或其他咽部成形手術前進行）

Speech Characteristics

Speech problems are not uncommon in children with cleft lip and/or palate. Cleft lip and/or palate has a negative impact on their speech. Some of them may have 'cleft palate speech' with atypical consonant productions, abnormal nasal resonance and abnormal nasal airflow. Speech errors may include pharyngeal stops, pharyngeal fricatives, nasal emission in pressure consonants and mid-dorsum palatal stops. Moreover, Hypernasality frequently occurs due to excessive nasal resonance. The typical speech characteristics associated with cleft lip and/or palate are listed as below:

1. Delay in speech sound development (pressure and non-pressure consonants)
2. Reduced speech intelligibility
3. Omissions and substitutions are more frequent than distortions
4. Glottal and pharyngeal substitutions or other 'backed' productions (e.g. alveolars to velars or glottals)
5. Nasal substitutions

Treatment and Management of Speech Disorders

Therapy approaches for speech sound disorders in children with cleft lip and/or palate include surgical intervention and speech therapy. Surgical intervention targets structural abnormalities and obligatory speech deviations secondary to velopharyngeal insufficiency. After surgical repair, speech therapy may be needed for children with cleft lip and/or palate who have developed compensatory speech errors. Listed below are common surgical interventions and techniques used in speech therapy:

A. Surgical Intervention

1. Repair of cleft lip (usually performed within the first 6 months of life)

6. 二次手術 修復上顎瘺管（如有症狀）
7. 牙槽植骨手術（由牙齒發育階段決定並與矯齒科醫生配合）
8. 改善牙齒咬合不正的正頜手術（例如：上顎前移手術—通常在青春期完成）(ASHA, 2009)

B. 言語治療

1. 引入輕柔的耳語或使用持續的 /h/ 來打破喉音模式，並在張開聲門下教授輕鬆的口腔氣流。
2. 在口腔塞音後插入 /h/ 以阻止在元音開始前使用喉塞音 [例如：p(h)op 表示 “pop”]。
3. 教授正確目標與補償性錯誤之間的聽覺辨別力，以便自我監察。
4. 提供視覺提示，包括：
   a. 使用側面圖或嘴巴模型作為視覺位置提示；
   b. 指向嘴唇 / 舌頭以提示位置
   c. 使用視覺氣流提示（例如：使用羽毛）；和
   d. 使用鏡子觀察發音位置——並排坐著，這樣孩子就可以看到他們自己和治療師的反映。
5. 使用語音位置技術，通常從雙唇音開始，然後移動到牙槽音。
6. 提供口頭提示，說明正確位置和方式或氣流等。
7. 提供觸覺提示，例如
   a. 感覺一個人的頸部肌肉組織，以幫助識別喉塞音、咽塞音和摩擦音的錯誤位置；
   b. 在發爆破音時，用手感覺噴出的空氣；
   c. 在發爆破音的過程中，用手指感受雙唇閉合和口腔氣壓；和
   d. 使用鼻塞 / 捏鼻（鼻堵塞）為個人提供口腔壓力的感覺並阻止錯誤的鼻腔氣流。
8. 使用既定的言語治療程序和技術（例如：最少對立體、傳統形式層次體統）。
   （Golding-Kushner，2001；Peterson-Falzone 等， 2016；Trost-Cardamone，2013）
9. 使用重建口部肌肉構音能力的提示 (PROMPT) 來塑造咬合架的運動並促進語音產生。
10. 使用口腔肌肉定位治療法（口肌工具）
11. 重度耳聾的人可以從視覺和觸覺反饋中受益，使鼻音正常化（Nguyen et al., 2008）。

2. Repair of cleft palate (usually by 12 months of age in a child with typical development)
3. Repair of submucous cleft palate
4. Procedures to correct noncleft velopharyngeal disorders or to correct postoperative velopharyngeal insufficiency (e.g., pharyngeal flap; sphincter pharyngoplasty; palatal lengthening procedures; posterior pharyngeal wall augmentation)
5. Tonsillectomy and/or adenoidectomy (may be needed prior to pharyngeal flap or other pharyngoplasty)
6. Secondary surgeries to repair palatal fistulae (if symptomatic)
7. Alveolar bone grafting (determined by stage of dental development and in collaboration with orthodontist)
8. Surgical treatment of malocclusion (e.g., maxillary advancement—typically completed in adolescence) (ASHA, 2009)

B. Speech Therapy

1. Introducing gentle whispering or using sustained /h/ to break the glottal pattern and to teach easy oral airflow with open glottis.
2. Inserting /h/ after oral stop consonants to discourage use of glottal stops prior to vowel onset [e.g., p(h)op for “pop”].
3. Teaching auditory discrimination between the correct target and the compensatory error to facilitate self-monitoring.
4. Providing visual cues, including
   a. using a lateral diagram or a model of the mouth for visual placement cues;
   b. pointing to lips/tongue to cue placement;
   c. using visual airflow cues (e.g., using a feather); and
   d. using mirrors for observing articulatory positioning—sit side-by-side, so that the child can view the reflection of themselves and the therapist.
5. Using phonetic placement techniques, typically starting with bilabials and then moving to alveolars.
6. Providing verbal cues such as instructions for correct placement and manner or airstream.
7. Providing tactile cues, such as
   a. feeling one's neck musculature to help identify incorrect placement for glottal stops and for pharyngeal stops and fricatives;
   b. feeling a released puff of air on one's hand during production of plosives;
   c. using one's finger to feel bilabial closure and oral air pressure on plosives; and
   d. using nose plugging/pinching (nasal occlusion) to provide the individual with the sensation of oral pressure and to discourage nasal airflow errors.
8. Using established speech therapy procedures and techniques (e.g., minimal pairs, traditional shaping hierarchy).
   (Golding-Kushner, 2001; Peterson-Falzone et al., 2016; Trost-Cardamone, 2013)

補償性言語錯誤的處理方法

治療方法：重建口部肌肉構音能力的提示法（PROMPT）

重建口部肌肉構音能力的提示法 (PROMPT) 是一種“實踐操作”的方法，它通過單詞和 / 或短語中的所有聲音，為服務使用者的口腔運動提供主動的觸覺 - 動覺 - 本體感受輸入，以提高語音排序的準確性。

為了提供與聲音的位置、方式和時間相關的發音提示，在嘴唇、面部、下巴和下頜舌骨上完成了不同類型的提示法。

咬合器的正確活動可以有系統地塑造。根據 PROMPT 運動語言等級 (MSH) 去選擇治療目標和進展。 MSH 分為 7 個語音子系統：

第一階段：音調；
第二階段：發聲控制；
第三階段：下頜控制；
第四階段：唇面部控制；
第五階段：語言控制；
第六階段：順序動作；
第七階段：韻律。

PROMPT 方法是由下而上的系統，強調下頜控制（第三階段），接著是唇面部控制（第四階段）和舌頭控制（第五階段）。

以下是 PROMPT 應用程序的一些示例：
1. 用食指和中指提供為 /p/ 的雙唇閉合觸覺提示
2. 於下頜舌骨肌的特定位置施加壓力，以提示軟顎塞音的正確位置。

9. Using the Prompts for Restructuring Oral Muscular Phonetic Targets (PROMPT) to shape the movement of articulators and facilitate speech production.

10. Using Oral Placement Therapy(TalkTools)

11. Individuals who are profoundly deaf may benefit from visual and tactile feedback to normalize hypernasal speech (Nguyen et al., 2008).

Treatment Approach for Compensatory Speech Errors

Treatment approach: Prompts for Restructuring Oral Muscular Phonetic Targets (PROMPT)

The PROMPT is a ‘hands-on’ approach which provides active tactile-kinesthetic-proprioceptive input to the clients’ oral motor across all sounds in a word and/or phrase to increase the accuracy of speech sequencing.

To provide cues related to the placement, manner and timing of a sound, different types of PROMPTS are completed on the lip, face, jaw and mylohyoid muscle.

Correct movements of articulators can be shaped systematically. Selection of the treatment goals and progression were determined following the PROMPT Motor Speech Hierarchy (MSH). The MSH is divided into 7 speech subsystems:

Stage I: tone;
Stage II: phonatory control;
Stage III: mandibular control;
Stage IV: labial-facial control;
Stage V: lingual control;
Stage VI: sequence movements;
Stage VII: prosody.

The PROMPT approach follows a bottom-up organization system where mandibular control (Stage III) is emphasized before labial-facial control (Stage IV) and lingual control (Stage V).

Below are some examples of PROMPT application:

1. using index finger and middle finger to provide tactile cue for bilabial closure of /p/
2. provides pressure on specific position of mylohyoid muscles for correct placement of velar stops

治療方法：口部肌肉定位療法（TalkTools 口肌工具）

口肌工具 (TalkTools) 是一種口部運動性肌肉定位療法，它通過使用一系列專門設計的工具結合了感官和觸覺導入。多感官方法包括一系列層次結構和程序，以發展和加強關鍵運動性技能，例如下巴穩定性、嘴唇閉合和舌頭抬高，以及提高餵食技能。

以下是 口肌工具 (TalkTools) 應用程序的一些示例：

1. 不同直徑和硬度的牙膠，以建立顎的強度和穩定性。
2. 壓舌棒和吹喇叭可促進唇部閉合和圓潤
3. 吸管增強下巴、嘴唇和舌頭之間的協調

用於鼻音過重和鼻腔排氣的生物反饋

生物反饋：鼻腔測量計 (Nasometer)

鼻腔測量計 (Nasometer) 廣泛用於評估與顎裂和其他顎咽疾病相關的鼻音。它為循證治療選擇，提供客觀測量和即時視覺和聽覺反饋。

生物反饋：鼻音氣流回饋學習器 (See-Scape)

鼻音氣流回饋學習器 (See-Scape) 可檢測說話時的鼻腔排氣。將尖端放入兒童的鼻孔時，任何氣體的排放都會令浮子在堅固的塑料管中上升，產生即時視覺反饋對氣流壓力和顎咽功能障礙的指示。這是測試氣流壓力而不是鼻音，表明顎咽功能缺乏而被視為鼻音過重，可用於治療和家庭練習。

Treatment Approach: Oral Placement Therapy(TalkTools)

TalkTools is an oral motor placement therapy approach which incorporates sensory and tactile input by using a range of specifically designed tools. The multi-sensory approach consists of a range of hierarchies and programmes to develop and strengthen key motor skills, such as jaw stability, lip closure and tongue elevation and also to improve feeding skills.

Below are some examples of TalkTools application:

1. Chewy tubes of different diameters and hardnesses to establish jaw strength and stability
2. Tongue depressor and horn blowing to facilitate lip closure and rounding
3. Straw to enhance association between jaw, lip and tongue

Biofeedback for Hypernasality and Nasal Emission

Biofeedback: Nasometer

Nasometer is widely used to assess nasalance associated with cleft palate and other velopharyngeal disorders. It provides objective measurement and immediate visual and auditory feedback for evidence-based treatment selection.

Biofeedback: See-Scape

See-Scape detects nasal emissions of air during speech. When a tip is placed in a child's nares, any emission of air causes the float to rise in the rigid plastic tube. Instant visual feedback and an indication of air-flow pressure and velopharyngeal dysfunction is generated. It tests air-flow pressure rather than nasality, which indicates a lack of velopharyngeal competence perceived as hypernasality. It can be used for both treatment and home practice.

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