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GHULAM QADIR FAYYAZ
EDITOR

Surgical Atlas of Cleft Palate and Palatal Fistulae

Springer

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With 1725 Figures and 50 Tables

Springer

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ISBN 978-981-15-8123-6 ISBN 978-981-15-8124-3 (eBook)
https://doi.org/10.1007/978-981-15-8124-3

This Springer imprint is published by the registered company Springer Nature Singapore Pte Ltd.
The registered company address is: 152 Beach Road, #21-01/04 Gateway East, Singapore 189721, Singapore

This book is Dedicated to

Intense hard days and long hours have been spent to produce the book , present in your hands, which I dedicate to multiple inspiring sources. Notable among them are

My father, Late Muhammad Siddique, who guided me to be a good human being,

My friend, Haji Muhammad Hanif Tayyab, who helped me to become a plastic surgeon,

My mentor, Late Prof. Kahlid Mehmood Durrani, who trained me in plastic surgery,

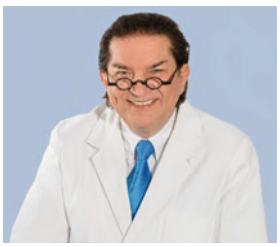
My Grand-Teacher in cleft craft, Late Prof. Samuel Noordhoff,

My patients who motivated me to do better and better every day of life, and lastly

My friends and family members who sacrificed their time and had faith in me.

Ghulam Qadir Fayyaz

Foreword



Prof. Dr. Hermann F. Sailer, MD, DDS
Dr. med., Dr. h.c. mult., Dr. sc. h.c. mult.
FDSRCS (Engl.), FRCS (Edinb.)
Founding President Cleft-Children International CCI
Chairman Sailer Clinic, Zurich, Switzerland

Cleft lip and palate is one of the major craniofacial anomalies and is prevalent worldwide – its incidence varies from 1:550 to 1:1,500 live births around the globe. Management of cleft lip and palate patients needs comprehensive cleft care programs, which can only be run in well-organized hospital settings. As most of the workload involves cleft patients in South Asia and the Pacific, Africa, and South America, government hospitals in these regions are unable to cater to such a huge patient workload. The majority of the population in these regions cannot afford the cost of management of cleft lip and palate patients in private hospitals. Even the private hospitals do not have all the facilities needed to help these cleft patients from birth to adulthood.

Charity organizations around the world have helped communities in underprivileged countries to provide free cleft surgery, as well as associated ancillary services such as nasoalveolar molding, cleft orthodontics, and speech therapy. Cleft-Children International (CCI), Switzerland, was founded in 2001 with the same initiative to help poor cleft patients in many parts of the world under a well-designed and guided program. CCI has always emphasized the multidisciplinary management of cleft lip and palate patients under compassionate, consistent, and well-organized services in most of the major cities.

I met with Prof. Ghulam Qadir Fayyaz in mid-May 2013, when he visited the office of Cleft-Children International CCI at Zurich. He asked me to make his organization CLAPP (Cleft Lip & Palate Association of Pakistan) a partner of CCI. As chairperson of CCI Foundation, I am always very eager to look into the management of cleft patients provided by our partners around the globe. Over the years, I have seen Dr. Fayyaz's commitment to help his cleft patients in a very comprehensive and integrated way.

Dr. Fayyaz's hospital is one of the most active cleft centers in the world. His team operates on 2,500 to 3,000 cleft patients every year, in partnership with CCI. He has extensive experience in the management of both primary and previously operated cleft lip and palate

patients. Fortunately, he is quite active in documenting his results for publications. His papers have won various international awards as well.

Dr. Fayyaz and his team have operated on more than 43,500 cleft patients so far and have very extensive experience in managing difficult and complicated cases of palatal fistulae. He has shared many of the palatal fistula patients with me that could not fit into any of the presently used classification schemes for palatal fistula. Accordingly, he has developed and published a new classification of palatal fistulae and dehiscence, and also an algorithm for managing these.

I am very happy that one of CCI's partner surgeons, Dr. Fayyaz, is writing this book, *Surgical Atlas of Cleft Palate and Palatal Fistulae*. As this is the first book that comprehensively covers different types of techniques for repair of cleft palate as well as palatal fistulae and dehiscence, this publication will become an ideal reference for all difficult and complicated cleft palate cases. His classification of palatal fistulae into midline, lateral, and subtotal cases is very helpful for designing a workable management plan. He has given us a practical and reproducible algorithm for the management of these fistulae. Each clinical chapter is dedicated to a specific type of problem, describes a management plan, and then provides step-by-step pictures during surgery with late postoperative follow-up.

I would be very keen to see this book published at the earliest opportunity so that cleft surgeons around the world will be able to help all their difficult palate patients in a much better way.

William P. Magee Jr., DDS, MD
Co-founder and CEO, Operation Smile Global Headquarters
Norfolk, Virginia, USA

What began back in 1982 as an opportunity for my wife and myself, along with the oldest of our five children, to take care of children with cleft lips and cleft palates in the Philippines has now evolved into one of the largest volunteer-based nonprofit organizations today. This would not have been possible without the thousands of good-hearted human beings who also recognized the opportunity that existed to help children around the world. Today, over 6,000 enthusiastic volunteers are willing to travel the globe and spend countless hours sharing their skills and talents with others.

From the beginning, we realized that education and training would be critical if our work was to be sustainable. We also understood that surgery is certainly not just the surgeon, but the cumulative knowledge of anesthesiologists, nurses, pediatricians, speech pathologists, child psychologists, dentists, and nutritionists, in addition to many other allied specialties. Change never occurs because of one individual or one particular specialty, yet it is the merging of these talents that is crucial.

We also realized early on that the need for cleft care is significant and we needed to enable a global network of talented individuals in order to reach more people earlier in their lives. Using

an integrated team approach to help improve the health and dignity of patients with cleft conditions in underserved areas, our dedicated volunteers have catalyzed the growth of Operation Smile over the years and improved our ability to provide comprehensive cleft care, while developing safe and effective surgical standards, thereby elevating the surgical infrastructure in the countries we serve. This enables patients to begin a new life with a lifted outlook from within. This also elevates the attitude of surrounding family and other members of their society.

By sharing our time, talent, and treasures, we not only create a lasting change for the child and their family, but also for society as a whole. One cleft surgery can transform a child's life in as little as 45 min, allowing them to breathe better, eat, speak, and live a life of greater quality and confidence. Operation Smile is committed to providing patients with health that lasts by being there to offer additional comprehensive services when needed. Thirty-one comprehensive cleft care centers in 19 countries across the globe now provide year-round medical services.

In taking care of the cleft child, there is always the possibility that a fistula may develop after the palate is repaired. This can significantly affect the functional results associated with cleft surgery. Management of the fistula is quite difficult, and excellent secondary care is essential. This requires careful planning and good execution to produce the desired results.

Up until now, books providing guidelines and management of palatal fistulae issues were limited. The knowledge, talent, and determination Prof. Ghulam Qadir Fayyaz has put into his book titled *Surgical Atlas of Cleft Palate and Palatal Fistulae* are remarkable. He has been an ally of Operation Smile since 2013, when we first met at a cleft congress in Orlando, Florida.

Since then, he has volunteered with Operation Smile on many missions all over the world, setting a positive example for others. His expertise in managing difficult palatal fistulae adds a wonderful dimension not only to international missions but to the organization as a whole, and I am certain his book will be a valuable asset for thousands of cleft surgeons, and their patients, around the globe for years to come.

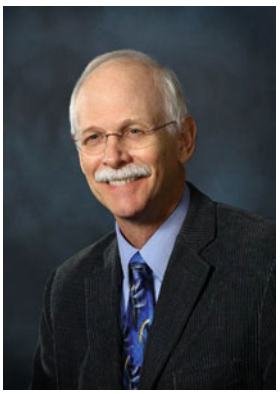
Joseph E. Losee, MD, FACS, FAAP
Dr. Ross H. Musgrave Endowed Chair in Pediatric Plastic Surgery
Associate Dean for Faculty Affairs, University of Pittsburgh School of Medicine
Professor and Executive Vice Chair, Department of Plastic Surgery
Division Chief, Pediatric Plastic Surgery, Children's Hospital of Pittsburgh of UPMC
Pittsburgh, Pennsylvania, USA

I was both thrilled and honored to be asked by Prof. Ghulam Qadir Fayyaz, from Lahore, Pakistan, to write a Foreword to his textbook titled *Surgical Atlas of Cleft Palate and Palatal Fistulae*. I was privileged to meet Prof. Fayyaz recently in July 2021 at the International Cleft Palate Master Course in Amsterdam (virtual) for the symposium "Palatal Fistulas: Prevention, Classification, and Treatment." As someone who has maintained a long-term interest in the

subject of palatoplasty, and specifically palatal fistulas, I, along with the other international experts in attendance, was nothing short of mesmerized by Prof. Fayyaz's presentations and overwhelmed by his experience treating palatal fistulae, which is likely one of the greatest – if not the greatest – in the world. He lectured on his incredible experience with successfully treating exceptionally complicated palatal fistulas, and, no doubt, this experience led to his classification system and successful therapeutic protocols.

This textbook, *Surgical Atlas of Cleft Palate and Palatal Fistulae*, is most certainly birthed from that massive personal experience, containing 109 chapters, 56 personally written by him. The remaining 53 chapters are penned by global experts from around the world. The exhaustive table of contents contains a comprehensive tour of palatoplasty from prenatal diagnosis to secondary surgery. All topics and procedures are covered, including alveolar, skeletal, and speech surgery, with chapters including wonderful illustrations and high-quality intraoral photographs.

I can personally attest to the massive effort of "birthing" such a work – to the sacrifice and dedication that it takes. It is nothing short of a labor of love – dedicated to those children born with orofacial clefts. For those of us who have committed our professional lives to the treatment of children born with cleft palate, this comprehensive encyclopedic tome of palatoplasty will benefit us all and be an invaluable resource and reference guide. We are indebted to Prof. Fayyaz for his significant contribution to our field.



Robert Mann
Associate Clinical Professor of Surgery and Pediatrics
College of Human Medicine, Michigan State University, USA

Greatness always requires inspiration.

For Prof. Fayyaz, inspiration came in the person of Prof. Ralph Blocksma, a plastic surgeon from Grand Rapids, Michigan, who moved to Pakistan in 1949, stayed there till 1954, and established United Christian Hospital in Lahore. Dr. Blocksma returned to Grand Rapids where he began a plastic surgery training program; the first trainee was Dr. Samuel Noordhoff, who in turn devoted his life in service of the people of Taiwan, where he established one of the most respected cleft programs in the world, another inspiration to Prof. Fayyaz. This was the center where Dr. Fayyaz was extensively trained in cleft surgeries at Chang Gung Memorial Hospital, Taiwan.

I was privileged to train with Dr. Blocksma and to count Dr. Noordhoff as my friend. When I first met Dr. Fayyaz, I saw a gentleman who was an extension of these two giants, not simply a surgeon, but a teacher and mentor, always going the extra mile to share his knowledge and skills.

Cleft care began with treatments for the adult cleft patients 150 years ago. These surgeries were deemed too risky for infants. Over the years, anesthesia improved and cleft care treatment was begun with progressively younger patients, finally including infants.

During this evolution, treatments that were developed for adults were used to treat younger and younger patients. We were taught to reconstruct the cleft palate defect by releasing tissue adjacent to the cleft, pulling the tissue together to close the gap, and using secondary healing to make up any tissue deficit. Because the infant face is immensely more compressible than the adult face, this seemed logical and more or less worked. Thus, for 15 decades, cleft palate care remained unchanged at its core.

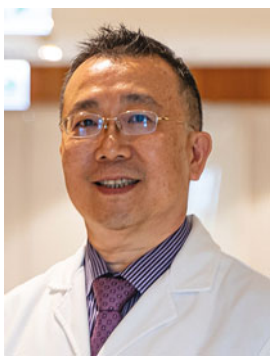
Over the many years, Prof. Fayyaz has treated some of the most complex problems associated with the cleft palate defect. Critically, he has also invested the time to document his work so it could be shared. All professions would be better off if the innovators documented and shared their learnings in this manner. Professor Fayyaz remains a willing teacher and, as with all great teachers, remains open minded to his own learning process.

This textbook is a vital collection of cases encompassing all aspects of cleft palate care. In each chapter the cases are analyzed, the problems studied to determine possible causation, and treatment plans established. It is said that textbooks are quick to be outdated, but the process of presurgical analysis so clearly documented by Prof. Fayyaz will stand the test of time. He gives us a fascinating look at the results of surgeries done using traditional repairs, including, but not limited to, infants. The book begins with the compressive (comprehensive) techniques used in treating the infant patient, moving on to describe other techniques used later in life to repair the resultant effects of undue facial compression. He includes compelling chapters on orthognathic surgery, which has been regularly required to repair midface hypoplasia.

But Prof. Fayyaz gives us so much more, as he extensively covers treatments for older children and adults. Studies of large tissue gaps closed in older patients using the traditional treatments give us an unprecedented look at the historic roots of the cleft repair philosophy, still very much dominant today. The amazing results that were achieved are a direct reflection of Dr. von Langenbeck's brilliant use of relaxing incisions and secondary healing. It was, of course, Dr. von Langenbeck who initially saw that the bones of the mature face had sufficient strength to hold a large space open, long enough for epithelialization to close the tissue deficit.

Thank you, Prof. Fayyaz, for putting together such a marvelous collection of cases. The question of the compressive power of healing by secondary intention in infants versus older patients has troubled cleft care practitioners for generations. This landmark book brings the answer alive before our eyes. And this is just one example of the myriad of issues waiting to be discovered with a deep dive into this eye-opening book.

This beautiful textbook is a reflection of Prof. Fayyaz, as well as his and my inspirations, Dr. Blocksma and Dr. Noordhoff. It offers a gateway to the future of cleft palate care.



Philip Kuo-Ting Chen, MD
Professor in Surgery, Taipei Medical University
Director, Craniofacial Center, Taipei Medical University Hospital, Taipei, Taiwan

Parents of cleft lip and palate babies usually pay more attention to the result of lip repair than palate repair simply because everyone can easily appreciate it. Moreover, if the change after lip

repair is dramatic, the result of palate repair can only be assessed after several years.

Likewise, cleft surgeons concentrate more on lip repair, and most of the textbooks on plastic surgery or cleft care provide more chapters on lip repair than palate repair, not to mention how few are those on the management of palatal fistula.

However, the result of cleft palate repair affects many areas of child development: speech, hearing, maxillary growth, and the process of swallowing. A poorly executed palate repair will have serious consequences, with the worst scenario of tissue necrosis and a huge fistula. It takes great efforts to reverse these complications, whereas only little can be found in the literature on their management.

Edited by Prof. Fayyaz, this book is a comprehensive textbook on all the knowledge about cleft palate repair, including primary and secondary treatment and postoperative care. Such detailed review on the classification and management of palatal fistula cannot be found in any other textbooks.

I have known Prof. Fayyaz since 2002 and have visited his centers in Faisalabad and Lahore many times. I have witnessed his effort to improve cleft care in Pakistan. I would like to thank him for his willingness to share his precious experience in cleft palate surgery and fistula management. This work is especially beneficial for surgeons in well-established centers who seldom have the chance to deal with these complications.

It is my honor to write this foreword, and I will recommend this book to all cleft surgeons around.

Mohamed El-Shazly, MBBCh, MSc, PhD, MD
Professor of Plastic Surgery, Assiut University, Egypt
Co-founder, CEO, Operation Smile, Egypt

What an amazing professional experience it is to become a physician and surgeon and what a glorious career to practice as a plastic and reconstructive surgeon! It is both rewarding yet demanding, interesting yet perplexing, and you should be willing to delve into the challenges of reconstructive surgery to complement aesthetic and cosmetic interventions. Profound changes in the landscape of healthcare and medical education are providing challenges as well as opportunities to the plastic surgery specialty to develop an international perspective and a global outreach and to address growing needs.

Cleft surgery is not a simple subspecialty. The techniques to repair and reconstruct the deformities are very complex, and the outcomes are both functional and cosmetic. It is also very critical because it is a matter of fate for the patients. Each cleft surgery means a different fate for the shape, the smile, the voice, the tone, and the consequent engagement in the community. It is not a “cut as you go” surgery. It is in your eyes, mind, and hands and through your scalpel that the fate is sealed. Please be careful.

I encourage all my colleagues at all levels of experience to be proud of our calling and our mission to serve as plastic and reconstructive surgeons. We practice the surgery for changing

shapes and changing lives. Do your best to master one or two techniques during your career learning from the masters. This would help improve your results and be better for your patients than trying to know and acquire all the techniques published and taught. Be keen to know more to ideally serve your patients. Include yourself in a cleft group or a charity cleft team, and volunteer your time and experience to help those patients in need and present them the humanitarian access to safe surgery.

Cleft surgeries are very crucial to the patients and their families, and they put their trust and fate in your hands and on your expertise. Always remember the feelings of a mother waiting 9 months and the disappointment she feels during the first ever look at the face and mouth of her baby. Always remember that it is not a usual surgery in your hands; it is a surgery of fate.

What a gift to work as a cleft surgeon and what a glorious opportunity to be included in a cleft team working side by side with others who share the common sense of helping people and serving patients with no attention to color, race, or religion. There is no other surgical discipline that combines a comprehensive care team spanning so many committed professionals. I was lucky to be a member of the Operation Smile team. I was fortunate to gain extensive experience and acquire knowledge through the years. I shared in the writing and editing of nine chapters in this distinguished book through my interest in cleft surgery, which I developed through this team.

Through the Operation Smile team, I met Dr. Ghulam Qadir Fayyaz, the editor of this invaluable book. A learned professor as well as a kind and gentleman! He dedicated his life to serve his mission and his patients. On a daily basis, he spends hours performing surgery, reading to enhance his knowledge, writing to help others learn from his experience, communicating with colleagues and students helping develop future scholars, and transferring his knowledge to others. A scholarly professor.

David K. Chong, MBBS, FRACS
Plastic and Reconstructive Surgeon
Royal Children's Hospital, Flemington Rd, Melbourne, Australia

I was having a chat with Billy Magee Jr. He has become a close friend. Close enough that we can disagree about things and still like each other easily. We were thinking for a term to describe surgeons who were leaders, who advocate for a region, and dedicate themselves to cleft care through education and service. These surgeons seemed to have a common denominator, being motivated by a currency that is not financial, and we wanted to coin a title to recognize their contribution.

Billy came up with the word champion.

I didn't like it.

I thought it sounded elitist. Could there possibly be these surgeons that are appropriately described in this way? I thought we could find a better name. Being stubborn, I mumbled that I thought there were other options. We wrestled awkwardly and then deferred the conversation.

As I begun to write this foreword, I got that familiar feeling of being wrong. I realized that there really are champions.

Because it describes my friend Ghulam Qadir Fayyaz perfectly.

How else could one describe a colleague who has done over 202 cleft missions and led a team that has operated on over 43,000 cleft patients in Pakistan and Afghanistan, patients who otherwise would have little access to care? The depth of his experience, his dedication to patients with clefts, and his willingness to share his techniques with others have left me in awe of him. When he told me he was going to do an atlas on palate surgery and fistulas, I was sure it would be done. I had previously been exposed to his passion and determination, and it has not surprised me to see him galvanize 38 authors (including many learned colleagues from his region) over the last 3 years to produce this atlas, as well as writing over half the chapters himself. Ghulam champions cleft care, his country, his colleagues, and education.

We know the best time to repair a palate is the first time. The problem of a poorly done primary repair sets the patient on a journey which is difficult to recover from. Even the small fistula is challenging, with its scarred tissue and propensity to recur, let alone the massive defects that Ghulam closes with his infectiously humorous demeanor, accompanied with straightforward instruction. I am sure you will share my opinion that his prowess dealing with large fistulas is unsurpassed. Thanks to this atlas, we all have a copy of his road map to some of the most difficult problems in cleft surgery.

Thank you for taking the time to gather these pearls for us, Ghulam. This atlas will fill a hole that is sorely required in cleft literature and will benefit many surgeons around the world and, most importantly, the patients they treat.

Oh, and I messaged my friend Billy to tell him that he was right. . . again.

Preface

The first ever cleft workshop I attended was in 2002 at Chang Gung Memorial Hospital, Linkou, Taiwan. I met legendary Prof. Noordhoff and Profs. Yu-ray Chen, Philip Chen, and Lun-jou Lo for the first time. It was a three-day workshop with lectures and live surgery sessions. On the last day, I asked Prof. Philip Chen about the concluding ceremony. "What? (as they don't have any formal concluding ceremony)", he responded. I told him that I would like to present some gifts from Pakistan to the faculty members of this workshop. He agreed to my request, and when the last lecture was finished, Prof. Philip Chen called me to the podium. I gave my camera to Prof. Myong Chul Park (from South Korea) to take pictures. I started as "Whenever anybody goes to a teacher (in the Indo-Pak region) to learn some art or procedure, they (as a tradition) usually take some gifts as a token of 'submission to the teacher' (the gifts may be a turban, a cap, or other items or some kilos of delicious sweet food items). I am presenting these gifts (on behalf of all the participants of this workshop) to the faculty members."

Instead of just three days at Linkou, Taiwan, I extended my stay and observed different cleft surgeries for an entire month. When I returned to Faisalabad, Pakistan, three new operating rooms (OR) at Punjab Medical College, Allied Hospital Faisalabad, were almost ready. I established these ORs by fundraising from the city of Faisalabad and made these functional by the end of 2002. In 2003, I founded a nonprofit organization, CLAPP (Cleft Lip & Palate Association of Pakistan), with the single purpose of providing free cleft surgeries to all deserving patients. Smile Train started supporting free surgeries (332 in 2003 and 617 in 2004).

On my promotion as Associate Professor of Plastic Surgery in 2005, I was transferred to the Services Institute of Medical Sciences, Lahore, and so the cleft program was shifted to Lahore as well. We established a small facility at Lahore in January 2008, dedicated to cleft lip and palate patients where we provide audiology, speech therapy, and cleft orthodontic services and cleft surgeries, totally free for all deserving patients. We were able to organize one of the busiest cleft programs in the world, where we operated 2800(2008), 3000(2009), 3600(2010) and 4222 (2011) cleft patients in a year's time. Our capacity building helped us to reach this level.

In 2004, we started cleft missions in remote areas of Pakistan, to provide free surgeries to the patients at their doorstep. We trained our surgeons, anesthesiologists, and supporting staff at our main center, CLAPP Hospital in Lahore, so that we could help hundreds of cleft patients in each of our cleft missions. In the first mission in 2004, we were two surgeons and two OR staff members; now we have grown to a 48-membered strong team. So far, we have conducted 193 cleft missions in Pakistan and Afghanistan. We published our experience in the paper "A Model Humanitarian Cleft Mission: 312 Cleft Surgeries in 7 Days" in *PRS Global Open* in 2015 (this paper won the Best International Collaboration Gold Award in 2016). Overall, CLAPP's team (since 2003 till date) has operated more than 43,500 cleft lip and palate patients.

Since 2013, we are working in collaboration with Cleft-Children International (CCI), Zurich, Switzerland. Most of my chapters describe the patients whose surgeries were sponsored by CCI.

We organized eight symposiums for the management of cleft lip and palate in Pakistan in 2004, 2005, 2007, 2009, 2011, 2013, 2015, and 2018. Professors Philip Chen and Lun-jou Lo were the most important faculty members and have contributed largely to the development of the cleft program in Pakistan.

Over the years, we gained extensive experience with handling cleft palate patients (and complications after palate surgery). All difficult palatal fistula patients from all over Pakistan and even Afghanistan ultimately landed in CLAPP Hospital, Lahore. We were able to innovate some methods to close difficult palatal fistula by simple techniques. We were operating more than 1500 cleft palate patients every year, both unoperated and previously operated patients (with fistula). We described our techniques of "Radical Dissection of Greater Palatine Artery" and "Continuous Versus Interrupted Sutures for Primary Cleft Palate Repair" (both papers were published in *PRS Global Open* and won awards). We tried to classify palatal fistulae as per prevalent classification systems; however, quite a good number of them could not be categorized properly. Over the years, we became more and more experienced and confident to handle a wide variety of palatal fistulae (not described previously), so we developed a new classification for them and devised an algorithm to close different and difficult palatal fistulae. We then Published our classification in PRS in January 2019.

Professor Philip Chen once told me that a cleft surgeon from the USA wanted to visit Taiwan for training in the management of palatal fistula. He told the American surgeon that we have extremely low fistula rate and even those are very minor fistulae. I then searched the Internet for any book on the management of palatal fistula, and to my utter surprise, I could not find even a single book solely focused on closure of palatal fistula. At that time, it occurred to me that I should try to organize our extensive experience on closure of different types of palatal fistulae into a book. Although the initial planning of the book was done in 2016, the overwhelming response to our paper in PRS (January 2019) fueled our passion to write this book, and we thought to include everything related to the management of palatal fistula. When I was trying to compile the list of chapters for the management of palatal fistula, it came to my mind that we should also include chapters on different types of palate repair, where we can share (with the readers) how to do a good repair of cleft palate and thus avoid complications of palate repair. To build a good foundation for primary palate repair, I then included chapters on the anatomy and physiology of normal and cleft palate. Later, I included everything related to cleft palate, namely, chapters on the etiology, genetics, psychosocial aspects, and antenatal diagnosis of cleft palate. This was a huge undertaking on my part, and I could not do it single-handedly. I contacted many of my friends in Pakistan and abroad to contribute to this atlas. I was assisted by friends from the Cleft WhatsApp group, created by Edwar Alvarez from Ecuador. Many friends from Taiwan, India, the Philippines, Indonesia, Australia, Egypt, Ecuador, Nicaragua, Guatemala, Peru, Paraguay, Brazil, the UK, Netherlands, Italy, and the USA were very supportive and contributed 33 chapters. Our colleagues from the Pakistan Association of Plastic Surgeons and allied disciplines were very helpful and have contributed 20 chapters. The rest of the chapters were written by me. When I started to write the book, it was estimated to have around 50 chapters; later, the number kept on increasing, and finally the book has swollen to 109 chapters.

There are a good number of articles published on the management of palatal fistula. However, there are limitations for the research articles: there is a word limit, and the number of figures is also limited. The idea of writing this book was to provide readers a detailed account of the management of each and every type of palatal fistula. To organize data of hundreds of patients having palatal fistula into a book was an uphill task. We tried to include those cases for which we had pictures of each and every step of the procedure. We also tried our level best to include those cases where we had very late follow-up for at least 3–6 months. It is the late follow-up which confirms the validity of a certain approach or invalidity otherwise. We routinely take multiple pictures during surgery for every difficult and interesting case. It took many months to select the cases which can be included in this atlas. There were many cases for each type of palatal fistula, and we selected the case which can better help the reader to

understand the most appropriate procedure for a particular type of palatal fistula. Once the cases were selected, we arranged the pictures in a sequence, from preoperative, perioperative to postoperative, and lastly, good follow-up minimum of three months. Then I started writing legends/captions for each picture. The description of the entire scenario was written and relevant references added. We selected more than one cases for each type of palatal fistula or primary palate cases, and in the end, we finalized the better among those cases for publication in this book.

We contacted Springer Nature in September 2018 to publish this book, and after many months of negotiations, we entered into a formal contract. What should be the name of the book? Initially, it was named *Surgical Atlas of Secondary Repair of Cleft Palate*, but it did not reflect the contents of the book. Then, we changed it to *Surgical Atlas of Palate Repair*, but even then there was no mention of palatal fistulae. Finally, after many consultations and deliberations, it was titled *Surgical Atlas of Cleft Palate and Palatal Fistulae*. I am thankful to Naren Aggarwal of Springer India and many other persons who have contributed to the development of this book.

Writing a book has never been an easy job. Sometimes, we can make a list of topics for chapters in the book and ask many colleagues to write one or two chapters from the given list. If the editor has one or two chapters of his own, the job becomes quite easier and the whole book can be compiled in about 6 months or so. The scenario here was quite different. I had to write 56 chapters on my own. It took me quite a long time to write each chapter, read it, reread it, and then rewrite it. Many of the authors sent their chapters in early 2020, but I was unable to review and edit their chapters as I was busy with my own chapters. But all is well that ends well. I am very happy and satisfied that we were able to compile a book which will enable thousands of cleft surgeons around the globe to manage their patients amicably.

June 2022 Ghulam Qadir Fayyaz

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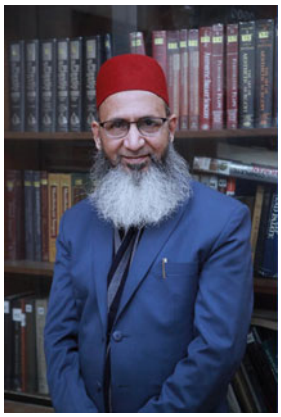

Ghulam Qadir Fayyaz Born in Multan in 1959, Ghulam Qadir Fayyaz graduated from Punjab Medical College, Faisalabad, in 1984. Then he completed postgraduate diploma in plastic surgery in 1988 from Vienna University, Austria. He obtained an MS (Master in Surgery) in plastic surgery from Dow Medical College, affiliated with the University of Karachi, in 1991.

He was appointed Assistant Professor Plastic Surgery at Punjab Medical College, Faisalabad, in 1995. He started the Department of Burns and Plastic Surgery at Allied Hospital, Faisalabad, in June 1999.

He raised funds from the business community of Faisalabad and installed multiple air conditioners in the Burn Unit to facilitate the burn patients. He raised funds from philanthropists of Faisalabad and abroad to construct three operating rooms (ORs) for the Department of Burns and Plastic Surgery. The three ORs were completed and equipped at a cost of 6 million Pakistani rupees. in 2002.

Dr. Fayyaz founded a nonprofit organization, Cleft Lip & Palate Association of Pakistan (CLAPP), in 2003, which was registered with the Government of Punjab, Pakistan.

The CLAPP Foundation was registered in New York in 2012, and the Internal Revenue Service granted income tax exemption for it in 2016.

Dr. Fayyaz was transferred from Punjab Medical College, Faisalabad, to the Services Institute of Medical Sciences, Lahore, and retired in January 2019 as Professor of Plastic Surgery from the same institution.

He was trained in cleft surgeries at Chang Gung Memorial Hospital, Linkou and Taoyuan, Taiwan. He has actively participated in cleft workshops arranged by Chang Gung Memorial Hospital, Taiwan, in 2002, 2004, 2006, 2008, 2010, 2012, 2014, 2016, 2019, and 2021. He was an integral part of cleft congresses held in Durban, South Africa (2005); Fortaleza, Brazil (2009); and Orlando, USA (2013).

Late Abdul Sattar Edhi donated 5 million Pakistani rupees to start a dedicated cleft hospital in Lahore, at 932-C, Faisal Town, Lahore. This hospital is the only cleft center in Pakistan where cleft lip and palate patients are provided cleft surgeries, audiology, cleft orthodontics, and speech therapy, all free under one roof. Since January 2008, this hospital has operated more than 15,000 cleft lip and palate patients from all across Pakistan and even from Afghanistan. Cleft surgeons from Germany, the UK, and Afghanistan have been trained in cleft surgeries at CLAPP Hospital, Lahore, in 2014, 2016, and 2019.

Over a period of 17 years, team CLAPP has conducted 193 surgical cleft missions – 187 in Pakistan and 6 in Kabul, Afghanistan. The team of Dr. Fayyaz has operated more than 21,600 cleft patients in these missions. Working since 2003, so far CLAPP has operated more than 44,000 cleft patients.

Team CLAPP has organized awareness seminars in collaboration with *Daily Jang* & Geo TV in 2010, 2014, 2016, and 2020. One seminar was conducted at Estiqlal Hospital, Kabul, Afghanistan, in May 2013.

Dr. Fayyaz's foundation, CLAPP, is going to start the construction of a 300-bedded charitable hospital at Main Ferozepur Road, Lahore.

He has written many research articles which have been published in local and international journals. Three of his papers received International Collaboration awards in 2016, 2018, and 2019 from the USA. He has written one chapter in *Global Cleft Care in Low-Resource Settings* published by Springer in 2021.

He is the chief editor of this book, titled *Surgical Atlas of Cleft Palate and Palatal Fistulae*, a 1,000-page book being published by Springer Nature. He has written 56 chapters, while 53 chapters have been written by other authors from around the globe.

He is the most sought-after speaker during international conferences on repair of palatal fistula.

He is an international volunteer surgeon for Operation Smile and has operated in Rwanda, Malawi, Egypt, Morocco, Paraguay, the Philippines, and Mexico.

He has organized eight symposiums in cleft lip and palate surgeries in 2004, 2005, 2007, 2009, 2011, 2013, 2015, and 2018 in Pakistan.

He has served as President-Elect of the Pakistan Association of Plastic Surgeons (PAPS) from 2018 to 2020 and President of PAPS from 2020 to 2022. Currently he is Chairman Board of Trustees of PAPS (2022-2024).

At above 60 years of age, he is still a force in the international arena of cleft care.

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Part I

Basics of Cleft Palate

Anatomy of the Normal Palate

1

Muhammad Ashraf Ganatra and Diaa Othman

Contents

Abstract

Craniofacial anomalies, in particular cleft lip and/or palate, are major human birth defects with a worldwide frequency of 1 in 700, and significant clinical sequelae. The normal palate is composed of the hard and soft palates and divides the oropharynx and nasopharynx. Development of the palate is the result of organized events at the cellular level, influenced by growth factors, receptors, and signaling pathways playing an important role in the growth, elevation, and fusion of the palatal shelves. Cleft palate results from failed midline fusion of these paired palatine shelves. Embryologic errors of formation leading to cleft palate include inadequate growth of the palatine shelves (e.g., failed neural crest cell migration), failed shelf elevation and fusion, and secondary degradation after fusion. This chapter reviews the normal palate development, with the relevant palatal embryology, anatomy, and physiology, with illustrative figures to enhance the understanding of its normal development and pathogenesis.

Introduction

Isolated cleft lip (CL) comprises about 25% of all clefts, isolated cleft palate (CP) is around 30% while combined cleft lip and palate (CL/P) cases account for about 45% of all cleft cases (Allam and Stone 2014). CL/P occurs more frequently and more severe in boys than in girls. Unilateral clefts are more common than bilateral clefts with a ratio of 4:1, and for unilateral clefts, about 70% occur on the left side of the face (Allam and Stone 2014). CL/P is frequently associated with other developmental abnormalities, and majority of cases are presented as part of a syndrome. Syndromic clefts account for about 50% of the total cases in some reports with about 300 syndromes described. Although the percentage of cases directly linked to genetic factors is estimated to be about 40%, all clefts appear to show a familial tendency. It

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G. Q. Fayyaz (ed.), *Surgical Atlas of Cleft Palate and Palatal Fistulae*,
https://doi.org/10.1007/978-981-15-8124-3_2

affects around 1.5 per 1,000 live births (about 220,000 new cases per year), with wide variation across geographic areas and ethnic groups, with substantial evidence of both health inequality and inequity (IPDTOC Working Group 2011; Mossey et al. 2011).

Anatomy

The palate forms the roof of the mouth and floor of the nasal cavities. It separates oral cavity from nasal cavity, paired maxillary sinuses as well as with nasopharynx. It consists of two portions, the hard palate in front, and the soft palate behind (Fig. 1).

The superior (nasal) surface of the palate is covered with respiratory mucosa, and the inferior (oral) surface is covered with oral mucosa, densely packed with glands. The normal palate divides the oropharynx and nasopharynx and is composed of the hard and soft palates (Allam and Stone 2014).

Embryology

Normal lip development occurs between weeks 4 and 8 of gestation. The mesoderm involved in facial development is believed to be a separate tissue similar to the primary germ cell types and of ectodermal origin. This specialized tissue begins to differentiate on or about day 21 of gestation as the ectoderm in the vicinity of the neural plate folds on itself to form the neural tube. Embryo is roughly 3 mm long.

As the neural tube takes shape, neural crest cells differentiate from the ectoderm and effectively separate the neuroectoderm of the neural tube from the covering cutaneous ectoderm. These special neural crest cells, although of ectodermal origin, exhibit most of the properties associated with mesenchyme, so that the tissue they form is termed "ectomesenchyme" (Fig. 2).

The normal hard palate is covered with a dense mucous membrane that adheres closely to the underlying periosteum, creating a mucoperiosteal covering of the oral bony surface (Aalst et al. 2008). The mucosa of the hard palate is tightly bound to the underlying bone; consequently, submucous injections here are extremely painful. The superior lingual gingiva, the part of the gingiva covering the lingual surface of the teeth and the alveolar process, is continuous with the mucosa of the palate; therefore, injection of an anesthetic agent into the gingiva of a tooth anesthetizes the adjacent palatal mucosa (Moore et al. 2014). Deep to the mucosa, are mucus-secreting palatine glands. The openings of the ducts of these glands give the palatine mucosa a pitted (orange-peel) appearance. In the midline, posterior to the maxillary incisor teeth, is the incisive papilla. This elevation of the mucosa lies directly anterior to the underlying incisive fossa. Radiating laterally from the incisive papilla, are several parallel transverse palatine folds or rugae. These folds assist with manipulation of food during mastication. Passing posteriorly in the midline of the palate from the incisive papilla is a narrow whitish streak, the palatine raphe. It may present as a ridge anteriorly and a groove posteriorly. The palatine raphe marks the site of fusion of the embryonic palatal processes (palatal shelves) which can be felt with the tongue (Moore et al. 2014).

The ectomesenchyme migrates along the natural cleavage planes between the mesoderm, ectoderm, and endoderm. In migration from their site of formation, the neural crest cells have no predetermined pattern of travel. The migration of this ectomesenchyme over and around the head is essential to the development of the facial processes (Fig. 2a, b, c, d, e, f). By the end of week 4, the frontonasal prominence is formed from migrating neural crest cells of the first pharyngeal arch. Nasal

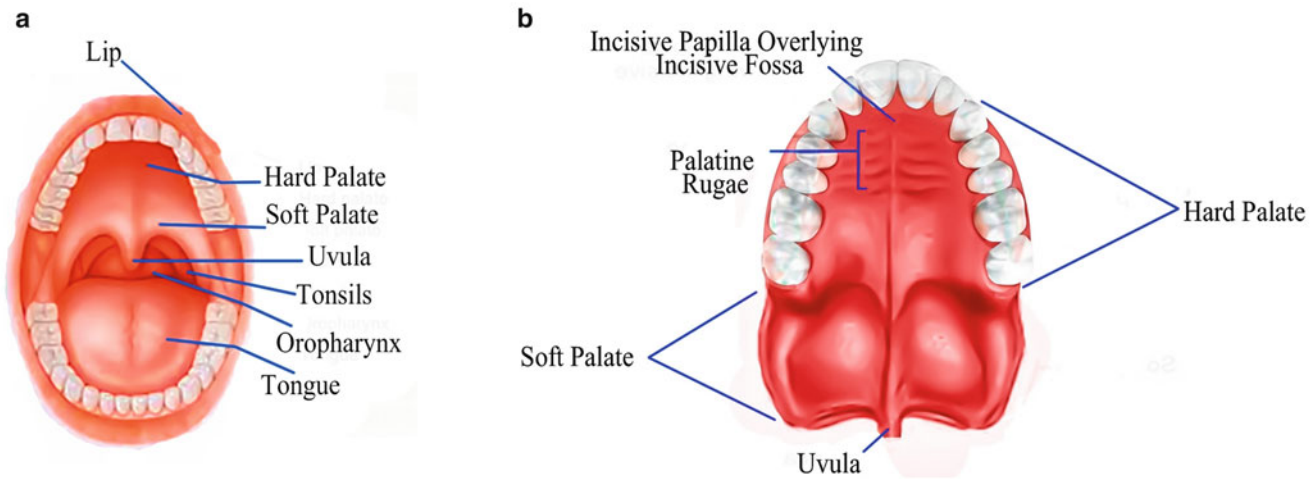

Fig. 1 (**a**, **b**) General outline of oral cavity

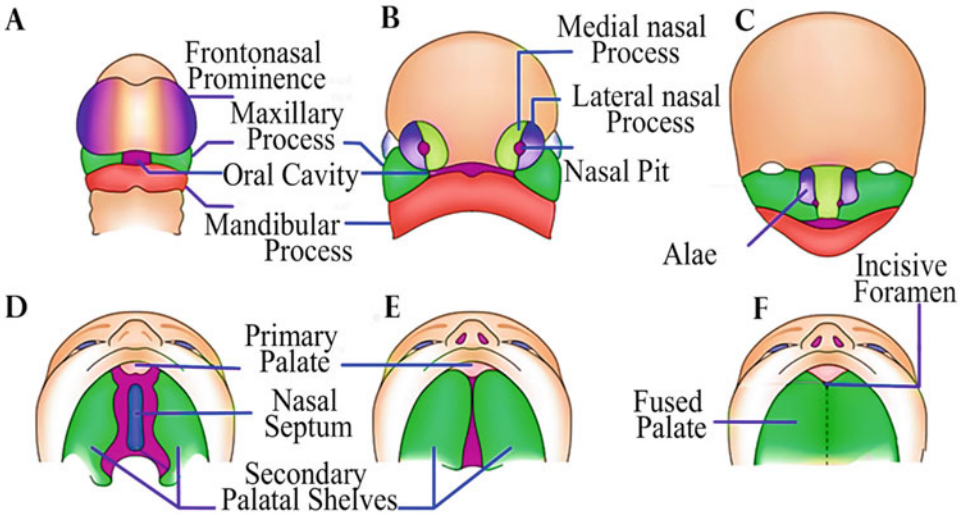

Fig. 2 (**a**–**f**) Different stages of development of palate

placodes, representing ectodermal thickening, develop at the caudal end of this structure and divide the paired medial and lateral nasal processes. The primary palate forms from the fusion of the paired medial nasal processes by week 6, giving rise to the premaxilla: central upper lip, maxillary alveolar arch and four incisor teeth, and hard palate anterior to the incisive foramen. The secondary palate develops after the primary palate during weeks 6–12. The medial projections of the maxillary processes form palatal shelves which rise above the tongue, fusing medially at the midline, anteriorly with the primary palate, and superiorly with the septum. The incisive foramen marks the anterior extent of the secondary palate. Formation of the primary and secondary palates completes the separation of nasal and oral cavities, permitting simultaneous respiration, and mastication.

Development

Normal development occurs sequentially; thus, cleft lip may or may not be associated with cleft palate. Similarly, isolated cleft palate may arise independently of cleft lip. Deformities of the lip, palate, and nose are a result of the disruption of normal development. The severity is dictated by the timing, the severity of the insult, and amount of disruption.

Anatomy of the Hard Palate

The hard palate maintains the width and anterior projection of the maxillofacial architecture, whereas the soft palate works as an active muscular valve, referred to as the velopharyngeal sphincter (Marks and Marks 1997; Aalst et al. 2008). This sphincter raises the soft palate toward the posterior pharyngeal wall, dynamically separating the nose from the mouth. The soft palate's intrinsic muscular function aids in proper breathing, swallowing, blowing, and phonation. Five pairs of muscles constitute the soft palate: the levator veli palatini (LVP), tensor veli palatini (TVP), mucularis uvulas (MU), palatopharyngeus (PP), and palatoglossus (PG) muscles (Strong and Buckmiller 2001; Aalst et al. 2008).

The primary palate includes all structures anterior to the incisive foramen (the premaxilla), whereas the secondary palate includes the hard palate, posterior to the incisive foramen, along with the soft palate. The muscular soft palate (velum) is found posterior to the hard palate (Friedman et al. 2005).

Hard palate is bounded in front and at the sides by the alveolar arches and gums; behind, it is continuous with the soft palate. It is concave in order to accommodate the tongue when at rest.

Hard palate is made up of two types of bones. Anterior two-thirds consist of palatine process of maxilla and posterior one-third consists of horizontal plate and pyramidal process of palatine bone, and they represent the posterior edge of the hard palate. Incisive foramen, greater palatine foramen, and lesser palatine foramen are present on the oral surface of the hard plate (Fig. 3).

Incisive foramen is a slight depression behind the central incisor teeth. Nasopalatine nerves pass from the nose into the palate from this foramen.

Greater palatine foramen is present medial to the third molar tooth at the lateral border of the hard palate. Greater palatine artery emerges from this foramen and is the main supply of mucoperiosteal flap raised for palate repair.

Lesser palatine foramen is posterior to greater palatine foramen and pierces the pyramidal process of palatine bone. These foramina transmit the lesser palatine nerves and vessels to the soft palate and adjacent structures (Moore et al. 2014).

Nerve supply of the palate is by greater palatine nerve which is a branch of third division of the trigeminal nerve,

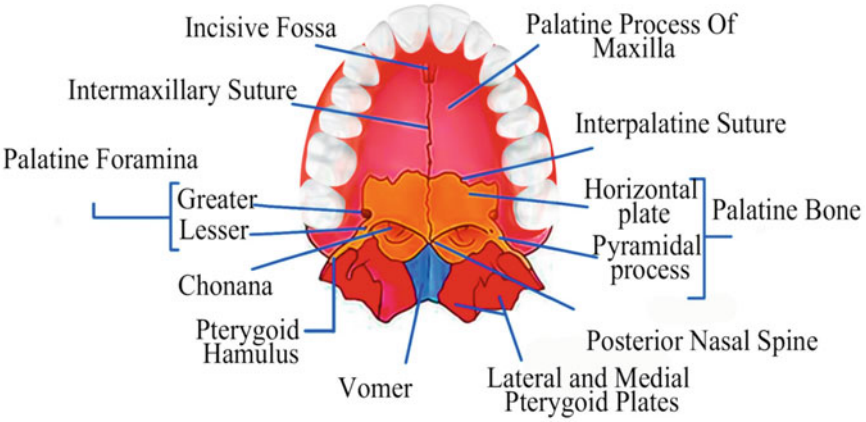

Fig. 3 Bony framework of the palate

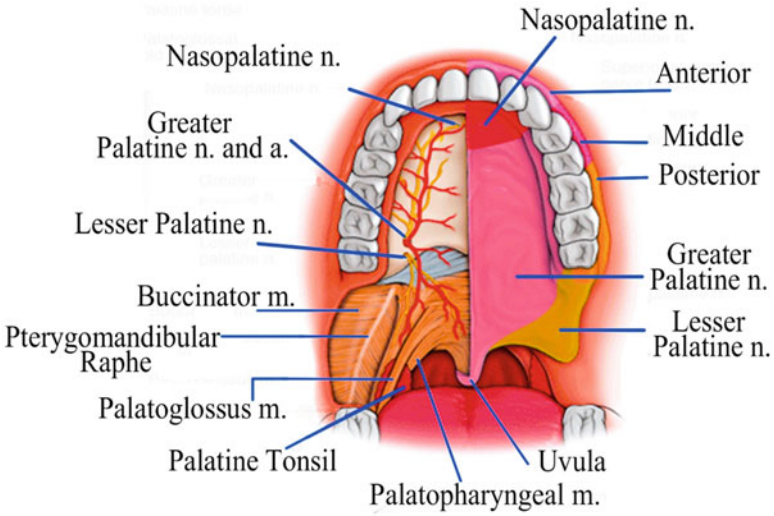

Fig. 4 Soft palate

that is, maxillary division. After arising from maxillary division, it passes through sphenopalatine ganglion and emerges with artery from greater palatine foramen.

Hard palate is covered by a dense structure, formed by the periosteum and mucous membrane of the mouth, which are intimately adherent. Along the middle line is a linear raphe. It is covered with stratified squamous epithelium, and furnished with numerous palatal glands, which lie between the mucous membrane and the surface of the bone.

Anatomy of the Soft Palate

The other name of soft palate is "Velum." The soft palate is a movable fold of posterior one-third of the palate, suspended from the posterior border of the hard palate. It forms an incomplete septum between the mouth and pharynx (Fig. 4).

Soft palate has no bony skeleton. It consists of two surfaces, oral and nasal. The mucous membrane on oral side consists of stratified squamous epithelium and on nasal side consists of columnar ciliated epithelium.

It is made up of anterior aponeurotic layer and posterior muscular layer. When occupying its usual position, i.e., relaxed and pendent (hanging down), its anterior surface is concave, continuous with the roof of the mouth, and marked by a median raphe. Its posterior surface is convex and continuous with the mucous membrane covering the floor of the nasal cavities. Its upper border is attached to the posterior margin of the hard palate, and its sides are blended with the pharynx. Its lower border is free.

The soft palate is the movable posterior third of the palate and is suspended from the posterior border of the hard palate. Its anterior aponeurotic part is strengthened by the palatine aponeurosis, which attaches to the posterior edge of the hard palate. The aponeurosis is thick anteriorly and thin posteriorly, where it blends with a posterior muscular part. Posteroinferiorly, hanging from the middle of its lower border is a small, conical, pendulous process called uvula, and arching