

Cleft Lip Management Protocol at the Royal Jordanian Rehabilitation Center: Four Years Retrospective Analysis

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ABSTRACT

Objective: Cleft lip is a common congenital anomaly in our population. It imposes serious psychological and social disturbances in both children and their families. Centralization of cleft lip care led to the great evolution of pre surgical orthodontist management and the continuous rejuvenation in surgical techniques, hence primary cleft lip repair with primary rhinoplasty has become the keystone of successful management. In this study we present our experience and current practice at the Royal Jordanian Rehabilitation center which were based on successful centralization of cleft lip care with strict commitment to rules of modern cleft multidisciplinary care.

Methods: This is a retrospective study of 100 patients with cleft lip/palate who were classified and surgically treated in our cleft unit from January of 2014 to January of 2018. Pre surgical orthopedics (nasopalveolar molding – NAM- and taping) were applied in 98% of the cases. Surgical technique included both Chang Gung cleft lip nasal repair (variant of rotation advancement techniques) and the Anatomic subunit approximation repair (variant of geometric repairs). Both Tajima semi open rhinoplasty and McComb techniques were utilized in primary nasal repair. All surgeries were performed under general endotracheal anesthesia on inpatient basis. All minor revisions for secondary deformities were performed on outpatient basis. Average follow up period is 2 years.

Results: The geographic distribution of deformity was highest in the north of Jordan followed by the middle and south (48%:44%:8% respectively). The age of the patients at the time of operation ranged between 3 And 6 months (mean age was 3.5 months). Male to female ratio was 72%:28%, ratio of cleft lip:cleft lip/palate (CL:CLP) was 52%:48%, ratio of unilateral to bilateral deformity was 80%:20%, ratio of complete to incomplete deformity was 56%:44%. In unilateral cases ratio of left to right deformity was 65%:35%. The average weight at time of surgery was 5.1 Kg. The most common associated congenital deformity was congenital heart disease (most common was atrial septal defects) in 25% of cases. Associated craniofacial deformities included: hypertelorism in two cases, binder syndrome in two cases, Van der Woude syndrome in one case, type 4 craniofacial cleft in one case and microtia in one case. 8% of cases had positive family history of cleft. Mean operative time was 120 minutes and mean hospitalization period was 36 hours. Morbidity consisted of 3 cases with stich sinuses and minor dehiscence and two cases of pneumonia. There was no mortality. The most common secondary deformities were vertical nostril collapse in 23% of the patients, hypertrophic scarring in 10% of the patients, excess vermilion deformity in 5%, wide prolabium in 2% and vermilion border mismatch in 2%.

Conclusion: Centralization of cleft lip care with multidisciplinary and inter disciplinary decisions between cleft team members led to the dramatic improvement in total care. Achieving near normalcy in the shape and function of the cleft patients is the definitive goal with decreasing the number and complexity of subsequent procedures.

Key words: Cleft lip, Cleft lip care, Cleft lip management protocol, Cleft lip repair

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Introduction

Cleft lip is a common congenital anomaly in our population.⁽¹⁾ It imposes serious psychological and social disturbances in both children and their families. With the great evolution of pre surgical orthodontist management and the continuous rejuvenation in surgical techniques, primary cleft lip repair with primary rhinoplasty has become the keystone of successful management. To achieve the definitive goals of cleft care (normal speech, near normal cosmetic appearance and occlusion), strict adherence to the modern cleft, interdisciplinary and multidisciplinary care must be set as role model.⁽²⁾In this study we present our experience and current practice at the Royal Jordanian Rehabilitation center which are based on successful centralization of cleft lip care with strict commitment to the rules of modern cleft multidisciplinary care.

Methods

This is a retrospective study of 100 patients with cleft lip/palate who were classified and surgically treated in our cleft unit from January of 2014 to January of 2018. Pre surgical orthopedics (NAM and taping) were applied in 98% of the cases. Surgical technique included both Chang Gung cleft lip nasal repair (variant of rotation advancement techniques) and the anatomic subunit approximation repair (variant of geometric repairs). Both Tajima semi open rhinoplasty and McComb techniques were utilized in primary nasal repair. All surgeries were performed under general endotracheal anesthesia on inpatient basis, mean operative time was 120 minutes and mean hospitalization period was 36 hours. In our postoperative care we routinely apply nasal stents immediately after operation and they are kept in place using taping for the first week. Breast or bottle feeding is resumed after full recovery from anesthesia, no restriction on baby hand movements and no arm restrains. Postoperative analgesia consists of paracetamol suppositories with intraoperative local nerve block. Three doses of intravenous antibiotics are given in the peri operative period then oral antibiotic is used in the first week. All of our patients are hospitalized for overnight observation. Most of patients are discharged at the first day after operation unless there is co morbid status necessitates more stay in hospital. At discharge from hospital full family education and instructions are given, the first appointment to clinic is given after one week, we use one of the smart phone applications as synergistic tool for communication with the family with standard frontal and basal view photography images captured by the family for any help or questions. Wound care involves taping with topical antibiotic ointment for the first ten days, baby bathing is allowed at the fourth post operative day, and no suture removal is required since absorbable sutures were used in closure (vicryl 7/0 simple continuous closure which usually falls down early when scar massaging ensues). Scar care is initiated at the tenth day post operatively using silicone gel based preparations with five minutes vertical massaging not less than five times daily. In cases of vertical height nostril collapse or nostril stenosis long term silicone nasal comforters and splints of different sizes are used for six months (Fig.8).

All minor revisions for secondary deformities were performed on outpatient basis. Average follow up period is two years. No exclusion criteria were set since all patients presented with cleft lip to our service were included.



Fig 8: long term silicone nasal stents are used in cases of nostril collapse or stenosis.

Results

The geographic distribution of deformity was highest in the north of Jordan followed by the middle and south (48%:44%:8% respectively) (Figuer.1) and this is a very interesting finding that needs further studies of environmental and genetic factors since the population of Jordan according to the department of statistics is distributed as follows: North 27%, central 63% and south 10%. The age of the patients at the time of operation ranged between three and six months (mean age was 3.5 months). Male to female ratio was 72%:28%, ratio of CL: CLP was 52%:48%, ratio of unilateral to bilateral deformity was 80%:20%, ratio of complete to incomplete deformity was 56%:44%. In unilateral cases ratio of left to right deformity was 65%:35% (Table I). The average weight at time of surgery was 5.1 Kg and the mean operative time was 120 minutes. The most common associated congenital deformity was congenital heart disease (most common was ASD) in 25% of cases. Associated craniofacial deformities included: hypertelorism in two cases, binder syndrome in two cases, Van der Woude syndrome in one case, type 4 craniofacial cleft in one case and microtia in one case. 8% of cases had positive family history of cleft. Morbidity consisted of three cases with stich sinuses and minor dehiscence and two cases of pneumonia. There was no mortality. The most common secondary deformities were vertical nostril collapse in 23% of the patients, hypertrophic scarring in 10% of the patients, excess vermilion deformity in 5%, wide prolabium in 2% and vermilion border mismatch in 2%. (Table II) Utilizing the Pennsylvania Lip and Nose Scoring System good results were achieved in 58% of the patients, fair results in 40% of the patients (all minor revisions were performed on outpatient basis) and poor results in 2% of the patients who required total revision. (Table I, II).

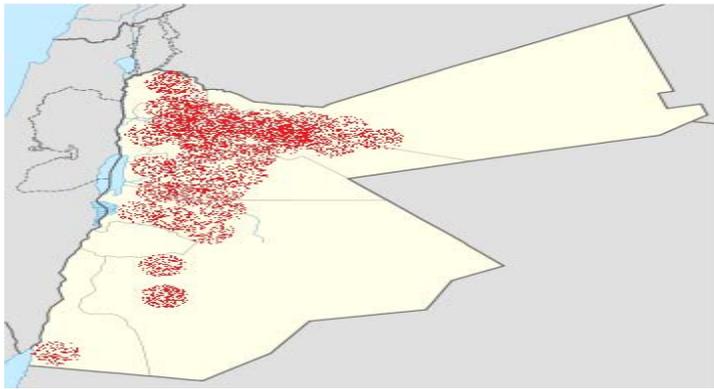


Fig1: geographic distribution of deformity was highest in the north of Jordan followed by the middle and south (48%:44%:8% respectively).

Table I: Patients' gender and cleft distribution

	% Of patients
Male	72%
Female	28%
CL	52%
CLP	48%
Unilateral	80%
Bilateral	20%
Complete	56%
Incomplete	44%

Table II:Distribution of secondary lip deformities (42%)

Secondary deformity	%from total number
Vertical height nostril collapse	23%
Hypertrophic scars	10%
Vermilion edge excess	5%
White roll/vermilion border mismatch	2%
Wide lip	2%

Discussion

The incidence of cleft lip/palate differs among races, and is approximately 1:1000 in those of Caucasian descent, 1:2000 of African, and 1:500 of Asian. In Jordan, Al Omari F¹ et al reported the overall prevalence rate for live births with cleft lip, cleft palate, or both was 1.39 per 1000 live births.

Cleft teams are comprised of experienced and qualified professionals; the key for successful management of cleft lip deformity is the multidisciplinary team approach and the active cooperation between plastic surgeons, orthodontists and other team members. This centralization in cleft care from interdisciplinary and multidisciplinary care point of view leads to sufficient numbers of patients each year to maintain clinical expertise in management.

Treatment plans are implemented on the basis of team recommendations with outcome measures and predictors to audit practice and monitor service quality (through studies) which lead to greater advancement in clinical research and evidence based practice with unified comprehensive database. ^(2,3)

With advances in prenatal diagnosis and ultrasonography; diagnosis of cleft lip is usually made after 16–20 weeks' gestation. Prenatal diagnosis gives time for family education, psychological support, investigation for other associated congenital anomalies and promising future for fetal surgery.⁽⁴⁾ A keystone of our cleft unit management target is the patient family. Cleft patients families are becoming an effective part of management and synergistic aid to cleft teams through full education, accurate information to families, realistic expectations and to understand subsequent steps in the complex lengthy surgical treatment plan. Definitive goals of cleft care are: normal speech, near normal aesthetic appearance and normal dental occlusion. Those goals are critical to improve function and eliminate psychosocial disturbances through optimizing the outcome while eliminating as many steps as possible in the treatment plan (Figure 2). ^(2,3)

Since best management for secondary deformities is prevention at the time of initial repair;the main aim of our practice is to achieve near-normacy in shape and function before the preschool age: before the age of individual social interaction in the community and before the age of face memory development in the child.(Figure 3) At initial presentation to our team, comprehensive family counseling is performed with detailed history and full family education regarding feeding and overview of timing for all steps required during the management course. Pediatric, cardiac, neurology, genetic and ENT consultations are taken. We encourage breast feeding if possible for isolated cleft lip and special feeding bottles (Haberman, Pigeon and Dr.Browns) for cases with cleft lip palate. Pre surgical orthopedics are very useful in the overall management especially in wide clefts to align lip/alveolar segments, reduce overall cleft width and improve cleft nasal deformity. This improves the overall function and aesthetics. Our routine practice includes passive devices (NAM and taping) applied as early as possible after the initial presentation till the time of definitive repair. ^(2,3,5) (Figure 4)



Fig 2: Pre and three years' post operative photos of bilateral cleft lip with satisfactory result.



Fig 3: four years follow up of unilateral cleft lip/palate patient, normal speech and function with near normal aesthetic result, minor secondary deformity of vertical height nostril collapse(for secondary septorhinoplasty at adult age).

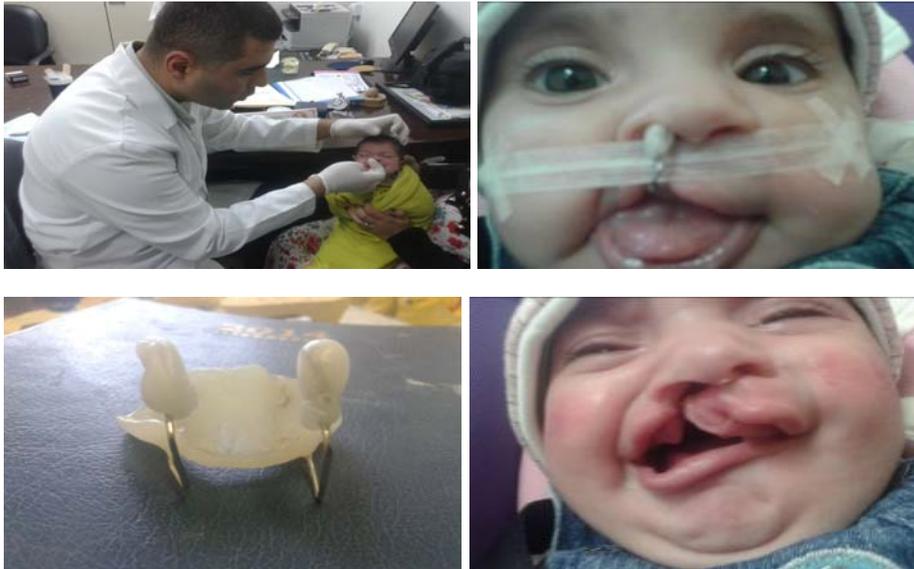


Fig 4: nasolabial molding NAM.

Unilateral cleft lip repair is often performed at three months of age, following the “rule of 10s”: 10 weeks, 10 g hemoglobin and 10 kg child’s weight. Several techniques have been described and all are variations of the Millard rotation-advancement technique or geometric repairs. Great rejuvenation in cleft lip repair surgical techniques has occurred over the last decade, since not all clefts are the same; cleft surgery has a lifelong learning curve and is a bag of tricks. With strict applications of general principles cleft surgeons are able to reach hybrid techniques which give the best results. Accurate application of full principles of anatomical and functional reconstruction at the time of primary repair aids greatly in minimizing the chance of secondary deformities development and decreases the number and complexity of subsequent procedures. ^(2, 3, 6, 7)

The key points in aesthetic, structural and functional cleft lip repair should apply the following main principles for the best outcome:

- Accurate examination of the deformity and pre surgical planing.
- Presurgical orthodontic molding of the alveolar segments, soft tissue and nasal molding to decrease the severity of primary deformity.
- Adequate release of malpositioned ala.
- Adequate rotation and release of abnormal orbicularis oris insertion on both sides of the cleft.
- Structural reconstruction of the nasal floor.
- Inter alar cinch stitch (using PDS 5/0 from the cleft ala to the non cleft side to control alar width) and septal anchoring suture of cephalic muscle (the abnormally released cephalic muscle is anchored to the caudal septum) for adequate advancement and to improve the overall nasolabial symmetry.
- Functional and structural reconstruction of orbicularis oris muscle with vertical or horizontal mattress sutures to create neo philtrum elevation.
- Adequate release of marginalis muscle and accurate positioning and use of Noordhoff triangular flap for vermilion reconstruction.
- Primary semi open rhinoplasty
- Tension free closure with meticulous skin care.^(6,7)

Many current techniques involve some component of early cleft nasal repair as evidence based in many studies; in our practice primary nasal repair has decreased social stigma signs in cleft nose and enhanced nasolabial harmony regardless if secondary septorhinoplasty is needed in future or not. ^(2, 3, 6, 7) In our practice we utilized both Chang Gung cleft lip nasal repair (variant of rotation advancement techniques) and the Anatomic subunit

approximation repair (variant of geometric repairs), both techniques yield satisfactory results. Both Tajima semi open rhinoplasty and McComb techniques were utilized in primary nasal repair, still long term studies are needed to evaluate and compare those different techniques in our practice. (Figure5, 6, 7)

In our post operative care we routinely apply nasal stents immediately after operation and they are kept in place using taping for the first week. Breast or bottle feeding is resumed after full recovery from anesthesia, no restriction on baby hand movements and no arm restrains. Post operative analgesia consists of paracetamol suppositories with intraoperative local nerve block. Three doses of intravenous antibiotics are given in the peri operative period then oral antibiotic is used in the first week. All of our patients are hospitalized for overnight observation. Most of patients are discharged at the first day after operation unless there is co morbid status necessitates more stay in hospital. At discharge from hospital full family education and instructions are given, the first appointment to clinic is given after one week, we use one of the smart phone applications as synergistic tool for communication with the family with standard frontal and basal view photography images captured by the family for any help or questions. Wound care involves taping with topical antibiotic ointment for the first ten days, baby bathing is allowed at the fourth post operative day, and no suture removal is required since absorbable sutures were used in closure. Scar care is initiated at the tenth day post operatively using silicone gel based preparations with five minutes vertical massaging not less than five times daily. In cases of vertical height nostril collapse or nostril stenosis long term silicone nasal comforters and splints of different sizes are used for six months. (Fig.8)



Fig 5: results of primary cleft lip nasal repair with the Chang Gung technique.



Fig 6: results of primary cleft lip nasal repair with the anatomical subunit approximation technique.



Fig 7: Tajima primary semi open rhinoplasty.

Despite this evolution in the overall cleft care secondary cleft lip deformities may persist especially in severe cases and cases of lack of family cooperation. These deformities involve: scarring, nasal collapse, vermillion border mismatch, long lip, short lip, vermillion excess and deficiency. The key for successful management of these deformities is prevention at the time of initial repair, for deformities that persist accurate diagnosis and knowledge of underlying etiology is the key in management. Fortunately most of our patient's secondary deformities were very minor and required minor revisions (Figure 9).^(8, 9, 10) Many virtual scoring systems have been proposed for aesthetic outcome evaluation of cleft lip repair results. In our study we utilized the Pennsylvania Lip and Nose Scoring System for the postoperative assessment of the result. This scoring system consists of two main parameters; it divides the nose into N1 to N3 and the lip into L1 to L3. N1, L1 account for almost perfect with nearly unnoted asymmetry at conversational distance. N2, L2 account for moderate deformities with some lip or nasal tip asymmetry at conversational distance and may require minor revision. N3, L3 account for severe deformities with significant lip and or nose a symmetry that will require a major revision (Figure 10).⁽¹¹⁾ Lip and nose score: Surgical outcome was good when the average score was one (no revision was necessary), fair if it was two (minor revision was indicated), and poor if it was three (complete revision of the surgery was deemed necessary). In this study group; good results were achieved in 58% of the patients, fair results in 40% of the patients (all minor revisions were performed on outpatient basis) and poor results in 2% of the patients who required total revision. Utilizing the Pennsylvania Lip and Nose Scoring System good results were achieved in 58% of the patients, fair results in 40% of the patients (all minor revisions were performed on outpatient basis) and poor results in 2% of the patients who required total revision. Most of the patients showed an acceptable vertical height of the nostrils although mild to moderate collapse was noted especially in cases of severe cleft lip-palate. Primary rhinoplasty is evidenced based in decreasing the overall cleft stigmata in childhood although dose not replaces the need for secondary rhinoplasty. (Figure 11)



Fig 9: early hypertrophic scar (arrow) and pulled up lip deformity due to scarring, accurate diagnosis is essential for management, since the patient had adequate rotation and harmony immediately after operation. This cicatricial pulled up lip deformity is successfully treated with silicone sheet, massaging and pressure taping.



Fig 10: white roll/vermillion border mismatch (arrows) which required minor revision with diamond shape excision and realignment.



Fig 11: Showing 4 years follow up in left cleft lip, patient underwent only minor revision for small excess vermillion deformity. Acceptable vertical height of nostrils seen from the primary rhinoplasty.

Limitations to this study were the lack of national registry of cleft patients, long term follow up is needed to conclude final results about the role of primary rhinoplasty and the possible need for secondary rhinoplasty. Furthermore long term double control studies are needed to compare and evaluate the results of the rotation-advancement versus the geometric repairs.

Conclusion

Centralization of cleft lip care with multidisciplinary and inter disciplinary decisions between cleft team members has led to the dramatic improvement in total care. Achieving near normalcy in shape and function of cleft patient is the definitive goal with decreasing the number and complexity of subsequent procedures.

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