THREE CASES OF COMPOUND COMPLICATED HARE-LIP,

OCCURRING IN THE SAME FAMILY; OPERATIONS FOR RESTORATION OF LIPS; WITH REMARKS ON THE OPERATION FOR CLEFT PALATE.

BY JAMES L. LITTLE, M.D.,

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XVIII. COMPOUND COMPLICATED HARELIP.

(CASES OF PROF. J. L. LITTLE.)

ARTOTYPE, E. BIERSTADT, N. Y.

THREE CASES OF COMPOUND COMPLICATED HARE-LIP,

OCCURRING IN THE SAME FAMILY; OPERATIONS FOR RESTORATION OF LIPS; WITH REMARKS ON THE OPERATION FOR CLEFT PALATE.

BY JAMES L. LITTLE, M.D.,

Professor of Clinical and Operative Surgery in the New York Post-Graduate Medical School, Professor of Surgery in the Medical Department of the University of Vermont; Surgeon to St. Luke's and St. Vincent's Hospitals, New York City, &c., &c.

The cases will be described in the order in which they came under my observation:

William Bocock, aged 21. John Bocock, aged 9. Charles Bocock, aged 18.

No hereditary tendency can be traced in father or mother's family. There were four boys and five girls. All the boys were born with hare-lip, while no deformity existed in any of the girls.

The order in which the children were born is as follows:

1. William. Compound complicated hare-lip. 2. A girl with no deformity.

3. Charles. Compound complicated hare-lip. A spindle-celled sarcoma made its appearance on the left side of the perineum in 1878, which I removed. It recurred, and I again removed it in October, 1882. 4. Girl with no deformity. 5. Girl with no deformity.

6. John. Compound complicated hare-lip; absence of ring-finger of right hand.

7. Girl with no deformity. 8. Girl with no deformity.

9. Boy with single hare-lip, who died in infancy.

These patients presented this deformity in almost the worst possible form, the arrest of development occurring at a very early period of fœtal life. The inter-maxillary bone in each case was distinct, being ununited to the superior maxillaries, and was continuous with the nasal septum and vomer. This projecting bone was partially covered by a tag of integument, which was continuous with that of the tip of the nose. In John (Case II), this bone contained two well-developed incisor teeth, while in William and Charles (Cases I and III), there was but one. There was a complete absence of both the hard and soft palate in all the cases, and in Case III the fissure was unusually wide (4 *cm.*). Articulation was so imperfect that they could be understood with the greatest difficulty.

Operations.—Case I.—William. Uranoplasty was performed at St. Luke's Hospital on February 9th, 1878. The sides of the fissure in the hard palate being imperfectly developed, and running obliquely upwards and inwards, the only operation that could be performed was the dissecting of the soft parts from the bone, from above downwards, and allowing them to meet at *B*, Fig. 113.

This was done, and there was sufficient material for the flaps to overlap in the central line. A small strip was removed from each flap and the edges accurately joined together. The union was complete throughout. The sutures were removed, and I felt sure of a satisfactory result. In about a week I found the line of union growing thin and showing indications of breaking away. An examination with the laryngeal mirror in the nasal cavity showed that granulations were springing up at the angles *c c*, and a line of ulceration was visible along the nasal surface of the wound, *B*. The union of the flaps finally gave way, and they assumed almost their original position in contact with the bony walls, *A A*, with the exception of a piece about one inch in width, at a point near the posterior edge of the hard palate. No further operation was permitted for the relief of this part of the deformity.

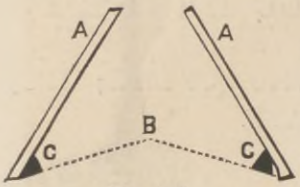

Fig. 113.

Operation for the Restoration of the Lip.—March 21st, 1878. The piece of integument covering the inter-maxillary bone was dissected up to the tip of the nose, and the projecting bone containing the incisor tooth was removed with bone forceps. The piece of integument which had been lifted up was then trimmed, turned down and accurately adjusted against the raw edge of the septum, so as to form a columna; this was retained in place by two silver wire and shot sutures, running directly through the septum. The union was perfect. On April 17th the following operation was performed:

The two portions of the lip were separated freely from the superior maxillary bones and a piece was removed on either side of the cleft by a pair of curved scissors. The posterior extremity of the newly formed columna was freshened, the edges of the fissure were brought together and retained in position by two pin sutures and a number of fine interrupted silk sutures. The columna was also nicely adjusted in the upper portion of the fissure. Good union took place everywhere except at the columna and the right side of the lip, and a considerable notch was also left in the centre of the lip. The boy went home during the summer to allow the parts to become soft and more yielding. He returned to the hospital December 9th, 1878. The edges of the opening between the columna and lip were freshened and closed with sutures. The union was perfect. On December 27th the final operation was performed on this case, and was for the purpose of relieving the notch at the line of union of the lip. Wharton Jones' operation, consisting of two incisions through the entire thickness of lip, on either side of the median line, extending from a point just below the columna downwards and outwards to but not through the vermilion border, forming an inverted V incision, was performed. The V-shaped flap included between these incisions was then pulled down, so that the notch was obliterated; the upper portion of the incision was closed by a pin suture, and the remaining portions by fine silk sutures. The lines of the incisions left after this operation was of the form of an inverted Y. The final result is illustrated in Plate XVIII, Case I.

Case II.—John, aged 9. The operation performed December 12, 1878, for the formation of a new columna, was the same as in Case No. I. On January 10th, 1879, the columna formed by the first operation being too prominent, its integument was dissected up and replaced after another portion of the bony septum had been removed, resulting in a well-formed columna. The operation on the lip was the same as in Case I, resulting in perfect union. Its appearance is shown in Plate XVIII, Case II.

In this case the vermilion border was perfect, but the lip was shortened so that the lower projected beyond the upper. In a few months the upper lip had stretched to such an extent that the difference was scarcely appreciable. This boy also had a very peculiar formation of the right hand, consisting in the absence of the ring-finger and its metacarpal bone. The little finger sprang off at right angles to the hand, just below the line of the carpal bones, and I cannot say whether it had a short metacarpal bone or not. It could be flexed in the manner shown in Fig. 114.

FIG. 114.

Case III.—Charles, aged 18. In this case the inter-maxillary bone was in contact with, although not united to, the right maxilla. The orifice of the right nostril was perfectly formed; the fissure in the lip and hard palate was unusually large, measuring four cm. in width. The mucous membrane covering the left inferior turbinated bone was hypertrophied, forming a prominent mass in the nostril. The operation was performed as in the preceding cases, with this exception: the columna being formed and the lips brought together at one operation. The union was perfect.

A supplementary operation was afterward performed. The left ala nasi being too widely separated from the septum, a V-shaped piece was removed between these two points, which were then brought together so as to form a perfectly oval nostril. A piece was also removed from the right side of the columna, reducing its thickness. The patient was discharged in the condition seen in Plate XVIII, Case III, still having a slight notch on the margin of the lip. Three years after, I found that all traces of the notch had disappeared.

In concluding this paper I desire to say a few words regarding Uranoplasty and Staphylorraphy. I had performed these operations successfully a number of times before operating upon the case described in the first part of this paper. Since that time I have carefully looked into the results, and find that although in a large proportion of the cases the operations are successful so far as the closure of the fissure in the hard and soft palate is concerned, yet so little, if any, benefit is obtained in the improvement of the articulation, that I have been forced to the conclusion that they should be discarded as surgical procedures in adults. I refer of course to cases in which the cleft is congenital. Mr. George Pollock says "the real object of the operation of closing the cleft in the palate is to enable the patient to articulate hereafter, plainly and intelligibly—*not* to enable the child to take food."* This last difficulty, he

* Holmes' Surgery, 2d Edition, Vol. IV. p. 423.

states, is overcome in a few days. My three patients had no trouble whatever in this respect. What the result would be if the operation was performed in early life, I have no means of knowing. From my own experience in operations upon the adult I can conclusively state that no improvement has ever taken place in the patient's articulation. The reason is undoubtedly this: the newly formed palate is rigid, tense, and deficient in length, and in a large majority of cases it cannot be brought into apposition to the pharyngeal wall, so as to close the buccal from the nasal cavities; and unless this be done perfect articulation becomes impossible. The division of the palatine muscles, which is necessary in the performance of this operation, also interferes, to a certain extent, with the proper use of the organs in speech.

While I have never seen a case in which the nasal twang was improved, I have seen a number of patients in whom an artificial palate rendered the articulation absolutely perfect. Norman W. Kingsley, M.D.S., D.D.S., of this city, who has paid a great deal of attention to this subject, has invented a soft artificial velum, which is so under the control of the surrounding and adjacent muscles, opening and closing the passages at will, that the wearer, after a certain amount of practice and education, acquires a perfect articulation. In one of my cases (No. III), Dr. Kingsley applied one of his artificial palates. Before introducing it an experiment was performed in my office, which I will describe in Dr. Kingsley's own words:

"Altogether the most extensive deformity of this kind, and the one having the most disastrous influence on the speech that I have ever seen, was a young man upon whom Professor J. L. Little, of the College of Physicians and Surgeons, New York, operated for compound hare-lip, and who afterwards came into my hands for an artificial palate. * * * Previous to the introduction of the artificial palate the following experiment was tried in the presence of a number of well-known surgeons: I wrote upon a slip of paper the following syllables, which the patient pronounced to the best of his ability, repeating each one several times: Bo, Lo, Ho, Mo, Ko, Po, Go, No, &c. The sound given by him to each of these syllables was written by the gentlemen present as nearly as they could be understood. A comparison of the various records showed that the only unmistakable syllables of the whole list were Ko, Go, and Ho, all throat sounds. Of the doubtful ones, No and Mo were interchangeable, and so were Lo and Ko; and of all the others no sound that he gave was any clew to the syllable he was trying to pronounce." *

The velum was applied, and the patient remained under Dr. Kingsley's instruction about two weeks only. He then returned to his home, where he was practiced in correct pronunciation by Dr. Levi W. Case. He returned, in three months, and was able to read a page from a medical journal before a number of physicians so that every word was understood.

I would urge, however, that the operation be performed if possible in early life, as recommended by Mr. Thomas Smith of St. Bartholomew's Hospital,† with the hope that, as the organs develop with the growth of the child, this difficulty will be overcome and artificial means dispensed with.

* A Treatise on Oral Deformities, p. 462. † Transactions Medico-Chirurgical Society, Vol. LI., p. 79.