REMARKS

ON

A CASE OF CONGENITAL WEB BETWEEN THE
VOCAL CORDS ASSOCIATED WITH
COLOBOMA OF THE LEFT
UPPER EYELID.

By SIR FELIX SEMON, M.D., F.R.C.P.

THE patient whose case forms the subject of the following communication was shown at the meeting of the Clinical Society of London on November 27th, 1885.¹ For some reason or other—speaking from memory after the lapse of so many years, I should say probably it was because I wished to append to my notes the result of the treatment then intended—I did not deliver my notes to the Secretary immediately after the meeting and the matter fell into oblivion. Looking through some old records, I accidentally came across my notes of the case not long ago; and as congenital diaphragms of the larynx are extremely rare, and the connection observed in this case of the laryngeal web with a malformation of the eye is, so far as I know, unique, I think it right to put the case, however late, on record.

B. S., a girl aged 16, applied at the Throat Department of St. Thomas's Hospital in the autumn of 1885, on account of hoarseness and occasional difficulty in breathing. She is one of a family of seven boys and girls, none of the others I was assured, showing any congenital deformity or deficiency. Immediately after her birth, a gap of the left upper eyelid throughout its entire length was observed. For about one or two years after birth she suffered from chronic incontinence of the bowels, which after that time spontaneously improved. From the moment of birth it was observed that the child's cry was both weak and hoarse, and, when the voice was used for crying, inspiratory stridor was observed. The stridor improved about the age of 7 or 8, but never entirely disappeared. It has in the course of the last few months again become worse. Her speaking voice has been very hoarse and weak throughout her life.

The eye mischief was under continuous observation at Guy's Hospital from one month after birth until three years afterwards, when Mr. Bader succeeded in closing the gap. Nothing has ever been done for the throat.

Present State.—The patient is a rather small otherwise well-formed girl. There is a deficiency of the central part of the eyebrows on each side, but much more developed on the left.

Eyes.—Right eye: she sees perfectly, slightly hypermetropic. Left eye: has a visual acuteness of one half when its hypermetropic astigmatism is corrected. (These data were kindly ascertained by Mr. Nettleship.)

Larynx otherwise well formed. Movements of vocal cords perfect. Between the anterior three-fourths of both vocal cords a perfectly symmetrical, somewhat transparent, slightly reddish triangular membrane is expanded, the free border of which is of a crescentic form, and apparently considerably thicker than the rest of the web; this border is of perfectly white colour. The glottic opening left between this crescentic border and the inter-arytenoid fold has the form of a horizontal-lying oval, and represents hardly one third of the width of the glottis under normal circumstances. The lateral borders of the web pass apparently over into the vocal cords without any appreciable limitation, the vocal cords being distinct from the diaphragm only by somewhat greater bulk and somewhat more rounded form. On attempted phonation the cords come

almost entirely together, and the web forms apparently a fold below their level. The voice is very hoarse, almost aphonic; after walking, speaking for some length of time, etc., inspiratory stridor occasionally is observed.

My notes end here. The following description of the treatment of the case is written from memory:

In view of the fact that during the last few months before the patient applied to the hospital there had been increasing dyspnoea, it was obvious that the condition could not be left to itself, and the question was only whether it was better to proceed by external operation, that is, thyrotomy, followed by cutting out the membrane with scissors, or by intralaryngeal operation. Of the former procedure I had had a discouraging experience. Once before I had removed a laryngeal web, the result of traumatism, by this method,² and although the operation in its immediate consequences had been successful, I had not been able, in spite of methodical

and long-continued dilatation, to prevent reformation of the diaphragm, which rendered the ultimate result of the operation quite nugatory. I therefore decided to give the intralaryngeal method a trial this time.

Originally it was intended to proceed as follows—namely, to insert Mackenzie's guarded laryngeal knife at the anterior commissure of the vocal cords close to the edge of the right vocal cord, and to cut through the web along that edge until close to the crescentic border of the diaphragm; then to repeat the same process along the border of the left vocal cord; and, finally, when merely a small bridge had been left, from which the triangular flap cut out would hang down, to grasp this bridge with cutting forceps and remove it bodily in the same way in which growths are removed from the vocal cords by intralaryngeal operation. I had not, however, sufficiently reckoned with the remarkable thickness and toughness of the web, factors which are insisted upon by almost everybody who has described cases of this sort. Nothing, as a rule, can be more fallacious than the appearance of such a diaphragm; it looks more or less transparent, and sometimes one thinks that one sees the rings of the trachea through it; but when it comes to the employment of slender cutting instruments, one too often finds that with all the force one can reasonably employ one does not make the least impression upon the tough structure, and a case is even on record³ in which the point of the knife simply broke during the operator's efforts to cut through the diaphragm. No such untoward accident occurred in the present case, but it was found simply impossible to incise, much less to cut through, the obstruction with either Mackenzie's guarded laryngeal knife, or later on with any of the stronger unprotected laryngeal knives that were employed. After many fruitless attempts, this method was finally given up, and the galvano-cautery substituted. This method, however, was not resorted to without serious misgivings; the breathing space left by the diaphragm was so small that it was obvious that if any considerable inflammatory reaction resulted from the employment of this method, very sudden and serious dyspnoea might occur. Under these circumstances, instead of availing myself of the galvano-caustic knife, I used a pointed galvano-cautery, and placed the point near the anterior commissure, in order to avoid as much as possible an extension of subsequent inflammatory reaction towards the small remaining glottic space. On establishing the current, the point of the galvano-cautery entered into the obstructing mass, but instead of quickly perforating it, as I had anticipated, with the hot point, I found I had entered into a dense mass of several millimetres thickness, through which the point of the instrument at white heat very slowly penetrated. Only when the platinum part, which was about 1 centimetre in length, of the instrument had almost disappeared in the mass, firm pressure being exercised all the time, it became noticeable by a sudden downward jerk of the instrument that at last it had pierced the entire thickness of the obstruction. It was quite difficult to withdraw the cautery through the narrow channel just made.

I distinctly remember that on this first occasion I did not repeat the puncture, as I wished to observe first whether any considerable inflammatory reaction would follow the galvano-caustic interference, and, secondly, whether there would be much tendency to a rapid closing up of the narrow channel made by the pointed instrument. In both respects I was most agreeably surprised when I saw the patient a few days later. There had been no reaction worth speaking of so far as could be ascertained, and the margins of the opening had remained as far apart as they were immediately after the operation. I then decided to repeat the perforation a number of times at some distance from one another, in the hope that the vitality of the web would thus be destroyed, and that it would ultimately be possible to get hold of its shrivelled remains by cutting forceps. The further steps of the galvano-caustic operation presented no difficulty whatever, and it was remarkable that the great thickness of the obstruction above referred to was limited to its anterior portion, and that towards the posterior free border it rapidly became much thinner. After a certain number of punctures had been made (I cannot say after so many years, and speaking from memory, how many altogether were required) the shrivelling process hoped for actually took

place, and this even in a much more satisfactory form than I could have expected. Seeing that the web apparently passed into the vocal cords without any definite border line, I had been much afraid that the ultimate result of my galvanocautic interference might have been that a number of shreds would remain hanging from the free borders of the vocal cords, which it would be very difficult to remove. But, contrary to this expectation, it was found that the shrivelling extended up to the vocal cords only, and there ceased abruptly, so that without any further operation the borders of the vocal cords gradually shaped themselves out quite nicely, and the voice became much stronger and more natural, whilst the dyspnoea entirely disappeared. The only part of the obstruction demanding further interference was the subglottic part on the anterior wall of the larynx, where the thickness of the tissue, as before stated, amounted to about a centimetre. These parts, however, could be removed by means of cutting laryngeal forceps, though a good deal of force had to be used in tearing out the tissue grasped between the branches of the instrument. It was quite remarkable how little reaction followed these operations, and, what was most important, there was no tendency whatever to renewed formation of the obstruction. Ultimately, practically the only sign of the mischief which had existed for so many years was a certain unevenness visible on laryngoscopic examination on the anterior wall of the larynx, below the level of the vocal cords. The voice had become quite loud, though remaining slightly veiled, and the difficulty of breathing of course had entirely disappeared. The patient attended off and on for some considerable time because I wished to be certain that no insidious reformation of the obstruction should take place. Ultimately I lost sight of her, but I remember distinctly that once again after a long interval she turned up simply to show herself, when it was found that the results obtained by the operation had been permanent.

REMARKS.

Membranous or pseudo-membranous webs between the vocal cords may be either acquired or congenital. In the first category syphilis and next to it traumatism are the most frequent causes of the condition. It is also said that "Stoerk's blenorrhoea" not seldom leads to the formation of membranous adhesions between the cords. Finally, there is a number of cases on record in which no definite etiology could be ascertained. Altogether these acquired diaphragms are rare, but not nearly so rare as those belonging to the congenital form. In his essay on *The Congenital Diaphragm of the Larynx*, published in 1893,⁴ Professor Paul Bruns enumerates 12 cases of this class altogether which he has found in the literature of the subject. He himself adds the thirteenth. Since then only two farther cases, so far as I have been able to find out, have been reported, one by Chiari,⁵ the other by Lacoarret,⁶ and the present case forms the sixteenth of the list. In all these congenital cases, with the sole exception of Chiari's, the aspect was curiously alike—that is, as in the case just described, the larger anterior part of the glottis was obstructed by an apparently membranous web connecting the anterior part of the borders of the vocal cords, and ending posteriorly in a semicrescentic ridge. In Chiari's case, however, a congenital web occupied the posterior part of the glottis. The thickness of the web in most of the cases was very considerable, and increased in the direction from behind forwards. The thickness near the anterior wall of the larynx in the case of Seifert and Hoffa above referred to, and in Bruns's own case amounted, like in my own just described, to 8–10–15 mm.

By far the most interesting series of these cases has been observed and described by Seifert. In his first communication on the subject⁷ he had affirmed, on the strength of statements made by the patient's relatives, that the girl came from a healthy family, in which no other malformations existed. Shortly afterwards he had to correct this statement⁸ after personal examination of the rest of the patient's family, when the following very remarkable state of things was disclosed:

1. The father, aged 48, quite healthy, with clear loud voice, has never suffered from hoarseness. Nevertheless, in the anterior commissure there is a membranous adhesion 3 to 4 mm. in width between the vocal cords, which, however, does not interfere with the movements of the cords.

2. The mother, aged 49, perfectly healthy, the larynx quite normal.
3. Son, aged 23. This young man was a half-brother of Professor Seifert's patient, her own father being his step-father. The larynx of this young man is absolutely normal.
4. The patient, whose case Professor Seifert had described, in which a large diaphragm, quite similar to the one described in my present case, had existed and had been operated upon.
5. A sister, aged 12. Speaks with natural voice, has never suffered from hoarseness or dyspnoea. In her larynx, however, a membranous adhesion of the anterior parts of the vocal cords exists, very similar to that seen in her father's case.
6. A sister, aged 7, born healthy, could cry loud, but from the very first some hoarseness in her vocalisation, to which not much attention was paid. There is some slight breathlessness on exertion. Voice somewhat hoarse but loud. On deep inspiration slight laryngeal stridor can be heard. In her case a membrane exists in the anterior part of the vocal cords, the size of which is about midway between that seen in Cases 1 and 5 on the one hand and that seen in Case 4 on the other. The membrane appears very thin, and on phonation it folds together below the vocal cords.

There could therefore be no doubt that the three girls had inherited the laryngeal anomaly from their father. This fact is particularly important, because the congenital nature of these webs has been doubted more than once. Seifert's observations establish this fact, I think, beyond doubt, and if any further corroboration were needed it is given by my own present case, in which the laryngeal anomaly was associated with so undoubted a congenital arrest of development as coloboma of the eyelid is. My case, so far as I know, is the only one on record in which the laryngeal anomaly in question was associated with some other congenital malformation.

The origin of the laryngeal malformation is still shrouded in considerable obscurity. Dr. Poore⁹ expressed himself as follows when bringing his case before the Laryngological Section of the London International Medical Congress:

"I formed the opinion that the web was congenital, first, because the patient's voice had always been peculiar since her first cry; secondly, because from her earliest infancy she has been liable to attacks of dyspnoea; thirdly, from the appearance of the web, which had nothing about it to suggest inflammatory or pathological origin. Two questions occurred to me. How were such webs caused? Either by the occurrence of disease or inflammation *in utero*, by the arrest of development, or the survival of some foetal condition. Normally, I believe, there never is a web in the larynx at any period of the development of the foetus, so that the last theory is untenable."

A series of sections of the larynx of human foetuses at various periods from the second month onwards, kindly presented to me by Professor A. Politzer, of Vienna, seemed to corroborate Dr. Poore's last statement; certainly in this series there is full patency of the glottis in its entire length throughout. But I find in Bruns's description¹⁰ an interesting statement bearing upon this question which points in the opposite direction. He discusses the explanation originally advanced by Seifert in his first case, namely, that the diaphragm was due to a hypertrophic inflammation of the vocal cords during intrauterine life, and rejects this as extremely improbable, seeing that the same malformation was present in three children of the same family (see above). "Possibly," he adds, "a phenomenon described by W. Roth¹¹ and observed in the course of the embryonic development of the larynx gives a clue. Roth showed that the upper part ("Anfangsstück") of the air tube in its first development is glued together, the epithelial glueing matter being formed in part from elements of the outer terminal layer (epiblast), and in part from the elements of the intestinal gland layer (hypoblast). The agglutination is not only found in the territory of the aditus ad laryngem, but it can be followed further down to a locality which corresponds to the rima glottidis."

"In an illustration given by Roth of a transverse section of the embryo of a rabbit of eighteen days, the rima glottidis throughout shows an agglutination in which the most superficial cells of the epithelial lining of the rima glottidis take part. In the hindermost part of the glottis a small gap can be seen, which Roth looks upon as the starting point for the later opening of the glottic space. This agglutinated mass, as shown by a series of sections, extends far down, and can still distinctly be recognised at the commencement of the trachea." Bruns continues: "Should those statements of Roth's be confirmed, the cause of the formation of diaphragms in the larynx would seem to be traceable to the agglutination of the

original formation, being only incompletely loosened and persisting in part. Slight indications of such membranes are often enough seen distinctly when making a laryngoscopic examination, a point already well insisted upon by Schrötter; sometimes the anterior commissure of the vocal cords, instead of forming an acute angle, represents a small bow line, sometimes below the commissure a slight fold of membrane extends from one side to the other, and projects more or less far into the character of the larynx."

The case now communicated would seem to lend some support to Roth's views, particularly so because, as in his series of sections, the agglutination was not limited to the vocal cords themselves, but in front extended almost a centimetre into the subglottic cavity. On the other hand it is difficult to explain Chiari's above mentioned case on the strength of Roth's statements.

The only question remaining is that of the treatment to be adopted in such cases. Undoubtedly in lighter cases—that is, such in which no respiratory troubles exist, and in which the voice is moderately or not at all affected, there may be no need to interfere in any way, and in a number of cases reported the policy of abstention has been followed. In cases, however, in which there is either total loss of voice or persistent or paroxysmal dyspnoea of a more serious character, or finally a combination of both aphonia and dyspnoea, surgical interference will be obviously required, and the only question is which treatment seems the most appropriate for these cases. To conclude from my own experience and from a study of the cases recorded in literature, I think that no hard and fast rule can be laid down. Much depends upon the thickness of the diaphragm, upon the patient's age and power of co-operation, and upon the tendency to re-adhesion during the process of removal. Theoretically one might think that thyrotomy, followed by total excision of the obstructing mass, would be both the quickest and the most radical treatment; but, apart from my own case above mentioned, Seifert and Hoffa state that they have had for a long time to carry on mechanical dilatation after the performance of thyrotomy and removal of the web in order to prevent readhesion which was threatening. On the other hand no definite rule can be laid down with regard to intralaryngeal treatment. In Krieg's case¹² mere incisions into the diaphragm produced such an amount of swelling that tracheotomy had to be performed. In Greenfield's¹³ and my own cases attempts at cutting the membrane with ordinary laryngeal knives were unsuccessful; in Seifert's case the knife even broke, as stated above. Bruns, to whom Krieg's patient applied after the performance of tracheotomy, succeeded by very persistent dilatation with special bougies introduced from the tracheal wound through the larynx in effecting complete disappearance of the laryngeal stenosis and restitution of normal voice. The treatment, however, carried on with interruptions, required a year and a half. Morell Mackenzie in his case obtained a good result by intralaryngeal removal of the web by means of cutting forceps. In my own case the combination of this method with the galvano-caustic treatment was equally successful. Should any future case come under my observation, I should make it entirely dependent upon the thickness of the mass which treatment to adopt, but should certainly give a trial to the intralaryngeal one before having recourse to thyrotomy.

NOTES AND REFERENCES.

¹ See programme of that meeting, BRITISH MEDICAL JOURNAL, November 21st, 1885, p. 1001. ² See BRITISH MEDICAL JOURNAL, 1878, vol. ii, p. 834. Meeting of the Royal Medical and Chirurgical Society, Case of Thyrotomy, etc.; and *Monats. f. Ohrenheilk.*, No. 6, 1879, Eine Thyreotomie. ³ *Berl. klin. Woch.*, 1886, No. 10. ⁴ *Archiv für Laryngologie und Rhinologie*, vol. i, Heft 1, 1893. ⁵ *Wiener klin. Woch.*, 1897, No. 25. ⁶ *Annales de la Polyclinique de Toulouse*, No. 5, Mai, 1897. ⁷ *Berl. klin. Woch.*, 1888, No. 10. ⁸ *Ibid.*, 1889, No. 2. ⁹ *Transactions of the International Medical Congress*, London, 1881, vol. iii, p. 316. ¹⁰ *Loc. cit.* ¹¹ *Mittheilungen aus dem embryologischen Institut zu Wien*, 2tes Heft, 1871, p. 155. ¹² *Atlas der Kehlkopfkrankheiten*, 1892, p. 112. ¹³ This case has, so far as I know, never been reported in full. Having seen it at St. Thomas's, I referred to it with Professor Greenfield's permission in my paper on Thyrotomy in the *Monats. f. Ohrenheilk.*, 1879, No. 6.

DONATIONS AND SUBSCRIPTIONS.—An anonymous donor has given £1,000 to endow a bed at St. Thomas's Hospital in memory of the late Charles Kingsley.—The Queen has forwarded her annual subscription of 100 guineas to the Middlesex Hospital.

CLINICAL LECTURE ON BRONCHO-PNEUMONIA, TO SHOW THAT PNEUMOCOCCAL PNEUMONIA IN A CHILD TAKES THE LOBULAR AND NOT THE LOBAR FORM.

Given at St. Bartholomew's Hospital,

By SAMUEL WEST, M.D.,

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I HAVE long been of opinion that the distinctions made between lobular pneumonia of children and the lobar pneumonia of adults do not hold with anything like the preciseness they should if the two conditions are really as distinct from one another as they are usually stated to be.

The question is largely a bacteriological one, and it is only recently that bacteriological investigation has turned in this direction.

I shall endeavour to prove that under the one term "broncho-pneumonia" several different conditions are included, and that many of them are pneumococcus inflammation; in other words, that some of the broncho-pneumonias of children are really the same disease as the common pneumonia of the adult, or, to put the proposition in another form, that the only difference between the pneumococcus inflammation of the adult and of the child is that in the one case the inflammatory consolidation which results is lobar or massive, and in the other lobular or patchy.

The evidence which I shall bring forward in proof of this thesis falls into two groups: the first clinical, the second bacteriological.

Of croupous pneumonia I do not intend to say much. The cases, as a whole, present a close family resemblance to one another; so much so that the less obvious, but not, therefore, less important, special features of individual cases are liable to be lost sight of. Yet it is clear on careful observation that there are many varieties of the disease, or many variations in it, and that these stand in close relation with the circumstances under which it has arisen, and with the course it is likely to run.

Bacteriology is gradually, step by step, explaining, or at any rate suggesting explanations of, these variations, and it is to this part of the subject only that I will briefly refer.

There is no doubt that the pathogenic organism with which croupous pneumonia is most commonly associated is the diplococcus pneumoniae or pneumococcus. But this is not the only pathogenic organism found, for the streptococcus, tubercle bacillus, Friedländer's bacillus, the staphylococcus and others have been present in different cases, sometimes alone, sometimes variously combined and associated. Still there is no doubt that the pneumococcus is by far the commonest of them all, for in about half the cases it exists alone, and in another quarter in combination with one or other of the bacilli mentioned. In the remaining cases other organisms are found, sometimes alone, more commonly in combination.

It thus appears:

1. That acute inflammation of the lung is not due to one pathogenic organism only, but to many and various organisms, though some are more likely to produce it than others, either because the lung is especially susceptible to their attack, or because they are more generally prevalent.

2. That the usual organism of croupous pneumonia is the pneumococcus.

3. That the different forms of, or variations in, pneumonia are very probably connected with different kinds of pathogenic organisms.

4. That even where the pneumococcus is the cause of the attack other pathogenic organisms may be associated with it, and thus produce variations in the symptoms; in other words, that the infection is not rarely a mixed one.

5. That secondary and subsequent infection with some other organism may account for a pneumococcus pneumonia ending in a particular way, as, for example, in abscess,