EFFECTS OF SULFONE TREATMENT ON THE LARYNX IN LEPROSY [1]

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In an article on tracheotomy in leprosy published in 1944 (1) I said, "Most lepromatous patients who live long enough develop laryngeal stenosis in time, and require tracheotomy." Experience with sulfone drugs since then has changed the entire picture. We no longer expect that most lepromatous patients will develop laryngeal stenosis; nor do we expect that this condition, when it does develop, will be irreversible. Patients who have been wearing tracheal tubes are removing them, while others have been able to avoid tracheotomy.

The use of sulfones at the Kalaupapa Settlement is relatively recent, although it was begun as soon as we could obtain them. We started promin in April 1946, diasone in February 1947, and promizole in October 1947. The figures in this paper are as of March 15, 1948, so the period covered is less than two years. In total, 116 patients are being treated with promin, 114 with diasone, and 15 with promizole.

CASES WITHOUT TRACHEOTOMY

Patients on "temporary release" because deemed no menace to the community, but remaining in the settlement by choice, are not discussed in this group. Of 284 "active" patients, 42 who have had tracheotomy at some time will be discussed later. Of the remainder, 45 have had no sulfone treatment. This leaves 197 patients never subjected to tracheotomy who have been receiving promin, diasone, or promizole. Of this group 152 give no history of laryngeal symptoms—24 of them were tuberculoid cases, in which one naturally would not expect involvement of the larynx—whereas 45 cases, all lepromatous, had experienced the cardinal symptoms of hoarseness or dyspnea, or both. No less than 37 of these 45 (82%) have improved with respect to this condition, apparently as a result of the treatment. Most of the 8 who did not improve complained of hoarseness rather than obstruction, and some of them have been under treatment only a relatively short time.

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[1] Read at the Fifth International Congress of Leprosy, Havana, April 3—11, 1948.

Of the 37 whose symptoms were improved, at least 2 were saved from operation by the treatment. Three more believe they would have required tracheotomy had the sulfones not been given, and probably there are others. Brief reports of the first two cases follow.

CASE 1. Hawaiian male, age 11. Admitted when 7 years old, with rapidly advancing lepromatous lesions. General health remained good, and in September 1946 treatment was started with pyricidin, of which he received 400 gm. in six months. During the last four months of this period he developed signs of laryngeal obstruction, with increasing dyspnea on mild exertion, so that it was feared he might require tracheotomy. Under hospital observation pyricidin was discontinued and treatment was begun with diasone, 0.15 gm. 3 times a week, gradually increasing to 0.6 gm. 6 times a week, with one week's rest in four. Dyspnea subsided rapidly, and in eight weeks he was permitted to resume normal activity. He has received 123 gm. of diasone in eleven months, with no serious toxic symptoms and with continued improvement in his general condition. It seems certain that the drug has saved him from tracheotomy.

CASE 2. Hawaiian-Caucasian male, age 38. Admitted three years ago with moderately advanced and progressive lepromatous lesions. During the first three months of 1947 he developed increasingly severe laryngeal dyspnea, which caused considerable concern because he persistently refused tracheotomy. He was employed as a cowboy, caring for the settlement's beef cattle, and we feared that fatal choking might occur when he was alone and far from help. He asked for promin but it was refused him, as we feared a flare-up of his laryngeal lesion. In March, I required him to stop work. Later, at his earnest solicitation and with much misgiving, I admitted him to the hospital for initiation of promin treatment. This was begun with 0.2 gm. daily, and gradually increased. In six weeks his dyspnea had cleared and he was able to resume work. His subsequent course has not been so satisfactory. He has had several bouts of lepra reaction, moderate hemolytic anemia, and a severe ulcerative lesion of his lower lip. Breathing remains good, however, and there is no doubt that promin has permitted him, at least for the present, to avoid tracheotomy.

TRACHEOTOMY CASES

A striking feature of our results is that, whereas formerly we had from 7 to 17 tracheotomies a year, the last one was in May 1947 (ten months ago). That patient had received only 170 gm. of promin before operation, and it was given irregularly because of cutaneous sensitivity. The preceding tracheotomy, performed in March, was on a patient who had received no sulfone before operation.

Of the 42 patients now in the settlement who have had tracheotomies, 3 had removed their tubes before sulfone treatment was started, and are not considered here. Of the others, 9 have been able to remove their tubes, presumably as a result of treatment. All of these had received promin, but 3 had been

changed to diasone because of poor veins. Of the other 30 patients 19 report marked improvement in breathing and voice. Some of them could now remove their tubes but do not do so because of nasal obstruction; they prefer the tube to constant mouth breathing. One patient breathes freely but is unable to speak above a whisper; the reason for his difficulty in phonation is as yet undetermined.

To the patients the most important thing is relief from dyspnea, especially from the "choking spells" which so often occurred to those who had worn tubes for a few years. Only one who has watched the fight for air of those whose bronchi are clogged by hardened crusts and swollen mucosa, fighting with them to relieve the obstructions and to give them a few more days of life, can appreciate the calm which has settled over the wards where the awful wheezing is no longer heard and the joy of not being required to write on death certificates, "chronic bronchitis due to indwelling tracheal tube." The last time I signed such a certificate was in February 1947, more than a year ago.

REMOVAL OF TRACHEAL TUBES

When a patient's condition has so far improved that removal of his tube seems indicated, we follow this procedure:

The voice is noted; it should be free, or nearly free, from hoarseness. The patient is instructed to hold his finger tightly over the tube opening and to inhale deeply; there should be little or no retraction at the suprasternal notch. An attempt is made to visualize the vocal cords by indirect (sometimes by direct) laryngoscopy; if the cords can be seen they should appear practically normal, be freely movable, and yield an adequate airway.

If the examination is satisfactory, we cover the opening of the inner tube with two thicknesses of adhesive tape. We have found this procedure preferable to using a stopper because pure natural rubber, the only safe material, has not been available. The patient is told to remove the entire inner tube quickly in case of dyspnea. After four to eight days, or longer if the patient has any doubt, and only if he is satisfied that he is breathing freely and may expect no further trouble, the tube is removed. A simple dressing is then applied, with transverse adhesive traction. In most cases the wound will close, leaving a deep dimple which eventually may flatten out. If the sinus is lined with epithelium, however, as may occur in cases of long standing, we operate when it seems probable that the opening will not close spontaneously.

The operation which we prefer is that of Jackson (2), with slight modifications. The skin is incised aroud the fistula, so that there will be a smooth closure with a vertical scar. The tract is then dissected free and the epithelium inside curetted at the base to promote adhesion. It is then put on tension and deeply ligated with chromic catgut; nonabsorbable sutures are not satisfactory because the lining of the tract is not sterile. The ligature pierces the fistulous tract twice, to produce firm anchorage and tight closure. Excess tissue is then removed, the stump touched with phenol and alcohol, and the skin closed with silk or dermal sutures.

OBJECTIVE FINDINGS

Laryngeal leprosy consists of lepromatous infiltration of all structures, plus edema and, at times, inflammation with production of adhesions. In severe cases the entire larynx may be so infiltrated and edematous that, even in a necropsy specimen, it is impossible to look inside until the larynx has been cut open. The epiglottis is usually involved and tends to be bent and greatly enlarged, so that it is difficult if not impossible to see into the larynx by either the indirect or the direct method. Stenosis occurs, and this is the reason for tracheotomy.

The degree of improvement which follows treatment will depend naturally on the severity of the original involvement, as well as on the duration and extent of treatment. Enlargement of permanently narrowed openings cannot be expected, although some relief may occur when edema subsides. In cases which have been less severely involved, especially where the infiltration has been largely nodular, the lesions may heal. Several of our patients have shown a remarkable absence of pathologic appearance following treatment. In almost every case, however, the epiglottis was distorted and enlarged, so that there were very few patients whose vocal cords could be examined with ease; in several instances our consultant found it impossible to see them.

In one patient the superior glottal opening was narrowed to a circular space about 3 mm. in diameter, yet beneath this opening the vocal cords appeared relatively normal. It is worth considering whether such a case might not benefit from surgery or dilation, with the tube still in place, after the active lepromatous condition has been quiescent for a sufficient period.

CONCLUSIONS

Improvement of laryngeal lesions is one of the most striking results of sulfone treatment, perhaps *the* most striking one. The reason for this is undetermined; possibly it is the generous vascular supply. It appears to make no difference whether the drug is promin or diasone, if it is used in adequate dosage over a sufficient period of time; and improvement may be seen in only a few weeks or months. We feel that patients who are beginning to show laryngeal dyspnea should be carefully watched for the first months of treatment, so that emergency tracheotomy may be performed if necessary; but often it may be avoided. It is our firm conviction that the response of laryngeal lesions to treatment would justify use of the sulfones even if no other benefit were derived from them.

SUMMARY

The sulfones—promin and diasone—are of great value in the care of patients suffering from lepromatous laryngeal involvment, both in those who have not required tracheotomy and in those who are wearing tracheal tubes. In an experience of less than two years with these drugs at the Kalaupapa Settlement, 9 patients have been able to remove tubes as a result of that treatment, and at least 2 (perhaps 5) have been saved from tracheotomy. The last death from the chronic bronchitis which follows tracheotomy was in February 1947; the last tracheotomy was in May 1947.

REFERENCES

1. SLOAN, N. R. Tracheotomy in leprosy. Internat. J. Leprosy **12** (1944) 11—30.
2. JACKSON, C. AND JACKSON, C. L. Diseases of the Nose, Throat, and Ear. W. B. Saunders Co., Philadelphia and London, 1st ed., pp. 497—8.